ISSN 1013 - 2295

Dhaka Shishu (Children) Hospital Journal

Vol. 34 | No. 2 | December 2018

Editorial

"Rationale for Adopting Pediatric Cardiac Program in Bangladesh"

Bangladesh Institute of Child Health

Dhaka Shishu (Children) Hospital

DHAKA SHISHU (CHILDREN) HOSPITAL JOURNAL

EDITORIAL BOARD

Chairman	:	National Professor Shahla Khatun, FRCS
Editor	:	Prof. Md. Jahangir Alam, FCPS
Executive Editor	:	Dr. Probir Kumar Sarkar, FCPS
Associate Editors	:	Prof. Shireen Afroaz, FRCP Dr. Mahfuza Shirin, FCPS Dr. Mustafa Mahbub, FCPS Dr. Maksudur Rahman, FCPS
Assistant Editors	:	Dr. Ipsita Biswas, MS Dr. Mohammad Abdullah Al Mamun, MD
Members	:	Prof. Waqar Ahmed Khan, M-phil Prof. Manzoor Hussain, FRCPCH Prof. Mohammed Hanif, FRCP Prof. MAK Azad Chowdhury, FRCP Prof. Samir Kumar Saha, Ph.D Prof. Abdur Rahed Khan, Ph.D Prof. M Kabirul Islam, MS Prof. Abu Ishaque Khan, DMRD Prof. Md. Abdul Aziz, MS Prof. Md. Selimuzzaman, FRCP Prof. Md. Monir Hossain, FRCPCH Prof. ASM Nawshad Uddin Ahmed, FRCP Prof. Syed Shafi Ahmed, Ph.D Prof. Md. Shahidul Islam, FCPS Prof. Farid Ahmed, MD Prof. Md. Mahbubul Hoque, FCPS Dr. Nilufa Akhter, M-phil

Published by Editor, Dhaka Shishu (Children) Hospital Journal, BICH & Dhaka Shishu (Children) Hospital, Sher-e-Bangla Nagar, Dhaka 1207. Tel: 9113048, 8122514, 9104211-20.
E-mail: info.dshjournal@gmail.com, website: dshjournal.org, www.bich.gov.bd

Published in May 2019

ISSN 1013 – 2295

Contents

Dhaka Shishu (Children) Hospital Journal

Vol. 34 No. 2 December 2018

Editorial

72 Rationale for Adopting Pediatric Cardiac Program in Bangladesh
Manzoor Hussain

Leading Article

74 Pneumonia: Post Hib and Pneumococcal Vaccination Era
Probir Kumar Sarkar, Samir K Saha

Original Articles

76 Immediate Outcome of Children Born with d-TGA: Experience in Dhaka Shishu Hospital Pediatric Cardiac Center
Abdul Jabbar, Nadia Binta Haque, Mohammad Abdullah Al Mamun, Manzoor Hussain, Rezoana Rima, Nowshika Sharmeen Echo

83 Iron Deficiency Anemia of Children Attending at Out Patient Department of Dhaka Shishu (Children) Hospital
Mahbubur Rahman, Rabi Biswas, Manzoor Hussain

88 Changes in Electrolyte in Different Stages of Perinatal Asphyxia: A Hospital Based Study
Sheikh Farjana Sonia, Soma Halder, Ruma Parvin, Azmeri Sultana, Shyla Rahman, Farhana Rahat

94 Effect of Topical Estrogen in Pre-pubertal Labial Adhesion
Gajendra Nath Mahato, Paritosh Kumar Palit, Md. Hasanuzzaman, Md. Moniruzzaman, Md. Abdus Sattar, Gouranga Kumar

98 Pattern of Pediatric & Neonatal Surgical Mortality: Experience at Dhaka Shishu (Children) Hospital
Md Samiul Hasan, KM Nur ul Ferdous, Umama Huq, Md. Abdul Aziz, Md. Ayub Ali, Ipsita Biswas

102 Pattern of AFP Cases Admitted in a Tertiary Care Hospital
Md. Rafiqul Islam, Mohammed Rizwanul Ahsan, Sabrina Makbul, Khaleda Islam, Rowshan Kabir, ABM Mahfuj Hassan Al Mamun

108 Pattern of Spirometry among Asthmatic Patients of Pediatric Age Group Who Attend the Outpatient's Clinic with Respiratory Symptoms
Md. Mosharaf Hossain, Md. Shakibur Rahman, Mohammed Rizwanul Ahsan, Md. Kamrunzzaman Kamrul

Review Article

112 Congenital Heart Block: A Review
Mohammad Abdullah Al Mamun, Rezoana Rima, Abdul Jabbar, Manzoor Hussain

119 Diagnosis and Management of Hirschsprung-Associated Enterocolitis
Md Ayub Ali, Md Samiul Hasan

Case Reports

123 Agenesis of Left Lung with Ostium Secundum ASD: A Rare Report
Nabila Akand, Probir Kumar Sarker, Md. Kamruzzaman Kamrul, Sakib Rana, Khondokar Ashiq-Ur-Zaman

127 Rare Presentation of Cystic Fibrosis Arthropathy- A Case Report
Khan Lamia Nahid, Md. Rukunuzzaman, Rana Kumar Biswas, Fahmida Begum, ASM Bazlul Karim

130 Gaucher Disease with Pregnancy - A Successful Outcome
Umme Parveen, Shahin R Chowdhury, Farid Ahmed

133 Abstract from Current Literature
135 Dhaka Shishu Hospital (DSH) News
136 Bangladesh Institute of Child Health (BICH) News
137 Postgraduate courses/training in paediatrics and child health
138 Students qualified from Bangladesh Institute of Child Health
139 Seminars, Symposiums, Workshop, CME / CPD
140 Instructions for Authors

DS (Child) H J 2018; 34(2) : 72-73

EDITORIAL

Rationale for Adopting Pediatric Cardiac Program in Bangladesh

Manzoor Hussain

There has been enormous success in prevention and control of vaccine preventable and infectious diseases worldwide including Bangladesh. Efforts at reducing mortality among children under 5 years of age have been the focus of significant international attention for decades and accelerated in 2000 with the Millennium Development Goals, with reduction of under-5 mortality rate by two-thirds. This work continued with the United Nations' adoption of the Sustainable Development Goals (SDGs) in 2015, specifically SDG 3, which aims to end preventable deaths of newborns and children under 5 years of age. Bangladesh is facing a multitude of health problems and congenital heart disease (CHD) is one of them. CHD occurs in 8 out of every 1000 live births. All countries including Bangladesh aimed to reduce neonatal mortality to at most 12 per 1000 live births and under-5 mortality to at most 25 per 1000 live births, though the rate of CHD remain constant.[1,2] So congenital malformations including CHD are now emerging as one of the leading cause of neonatal and under-5 mortality in Bangladesh. Without proper diagnosis and treatment, a majority of infants and children with cardiac disease both congenital and acquired die in developing countries and bear an increasing burden on health systems. Except congenital Rubella associated cardiac anomaly, CHDs are not preventable though, mortality could be brought down by programmed cardiac care and management.

In developed and developing countries there is wide gap regarding pediatric cardiac care. Absence of pediatric cardiac centers, presence of cardiac centers only in large cities and absence of specific health care policies in various countries are the reasons for this variation.[3] Scarcity of pediatric cardiac care in public hospitals, expense of treatment in private hospitals, lack of resources and trained personnelin this field and lack of awareness are the major reasons due to which treatment for CHD is currently out of reach for a majority of children in Bangladesh.[4] There are only few cardiac centers in the country scattered primarily in the capital city. Many of them are supported by adult cardiac program. Government institutions with their limitations of infrastructure, equipments and specialized support systems required cannot afford such facilities in large numbers.

The pediatric cardiac surgical program is rendered unviable due to economically weaker young couple at the start of their careers not able to afford the cost of the surgery and lack of government support. Realizing their inadequacy, many of the states in India, have chosen to support such patients through insurance schemes to facilitate surgeries at tertiary referral private hospitals.[5,6] A flagship scheme of Indian Government (Rashtriya BalSwasthya Karyakram [RBSK]) was launched in 2013 with a mandate to screen all children, aged 0-18 years for early detection and management of birth defects and other diseases.[7] There has been an overwhelming response to this initiative and has resulted in a sudden growth in the number of small and large cardiac centers in India.

The success of a pediatric cardiac surgery program is dependent on the development of a team of individuals comprising the pediatric cardiologist, the cardiac surgeon, intensivist, the pediatric cardiac anesthesiologist, perfusionists and nurses. Structured training programs are necessary for sustainability and growth of this subspecialty. China developed in-country training programs which have

Correspondence to: Prof. Manzoor Hussain, Professor & Head of Pediatric Medicine and Pediatric Cardiology, Bangladesh Institute of Child Health and Dhaka Shishu (Children) Hospital, Dhaka, Bangladesh. E-mail: manzoor.hussain717@gmail.com.

been shown to be feasible and cost-effective.[8] Realizing the importance of specific training in the field of pediatric cardiology, a Fellowship program by Bangladesh College of Physicians and Surgeons and MD Residency course in Pediatric cardiology under Bangabandhu Sheikh Mujib Medical University is initiated but no Pediatric cardiac surgery course have started yet.

Standalone pediatric cardiac hospitals tend to be very resource intensive. Piggybacking pediatric cardiac surgical programs on a successful ongoing adult cardiac program is practiced in many centers as it optimizes resource utilization. The cardiac catheterization lab, operating rooms and other lab services are shared for both pediatric and adult patients. In such 'adult-program-first' models, the pediatric cardiac program may gradually be expanded.

Government should stress the need to increase the capacity to care for children with heart disease by strengthening health systems, build a trained pediatric cardiac workforce to diagnose and treat children with heart disease and include pediatric cardiac care in benefits packages so that poverty will not be an obstruction.

References

1. UNDP Data.Retrieved May 17, 2017, from http:// www.undp.org/ content/undp/en/home/sustainable-development-goals/goal-3-goodhealth-and-well-being/targets.
2. Forouzanfar MH, Afshin A, Alexander LT. Global, regional, and national comparative risk assessment of 79 behavioral, environmental and occupational, and metabolic risks or clusters of risks, 1990-2015: a systematic analysis for the Global Burden of Disease Study 2015. *Lancet* 2016; **388**: 1659-24.
3. Pezzella AT. International Cardiac Surgery: A Global Perspective. *Seminars in Thoracic and CV Surgery* 2002; **14**:298-320.
4. Hussain M. Creating awareness for early identification of congenital heart disease. *Bangladesh Journal of Child Health* 2015; **39**: 1-5.
5. Rao SG. Twists, travails and triumph of pediatric and congenital heart surgery in India. *Ind J ThoracCardiovascSurg* 2006;**22**:108-
6. Nair SG. Pediatric cardiac program in India: Changing perspectives. *Annals of Cardiac Anaesthesia* 2011; **2**: 79-81.
7. Saxena A. Pediatric cardiac care in India: current status and the way forward. *Future Cardiol*2018; **14**(1): 1-4.
8. Pezzella AT. Cardiothoracic surgery residency program in Shanghai, China. *Ann. Afr. Chir. Thor. Cardiovasc 2009;* **4**: 81-99.

DS (Child) H J 2018; 34(2) : 74-75

LEADING ARTICLE

Pneumonia: Post Hib and Pneumococcal Vaccination Era

Probir Kumar Sarkar[1], Samir K Saha[2]

Pneumonia is the leading infectious cause of morbidity and mortality in young children of Bangladesh. It is endemic in large parts of the world and in particular, in developing countries, as well as in many indigenous communities within developed nations. *Streptococcus pneumoniae* (pneumococcus) and *Hemophilus Influenzae* type b (Hib) are the causative pathogens of serious childhood invasive disease, including meningitis, bacteremia and pneumonia. Both pneumococcus and Hib colonize the nasopharynx and spread through respiratory droplets. Children younger than 5 years are most susceptible to infection with the risk of infection greatest in children less than 2 years of age and *S pneumoniae* and *Hib* were the major causes of child mortality in developing countries.[1] Bangladesh has the fifth-highest rate of child pneumonia in the world, with an estimated 6 million cases annually among children under five.[2] In Bangladesh, 15% of all deaths among children under five are due to severe pneumonia.[3]

Conjugate vaccines against Hib have been used in the U.S., Europe and some other countries for almost three decades. They virtually eliminated Hib as a significant public health threat in areas of high and sustained coverage. However, children in several developing countries with high disease burdens have only started to receive Hib vaccine in the past decade. In 2009, vaccine against pneumococcus, known as the pneumococcal conjugate vaccine (PCV), began to be used in many low-income countries, where much of the disease burden is found. In Bangladesh, Hib vaccine was introduced in 2009 with significant reductions of both pneumonia and meningitis followed by 10 valent Pneumococcal Vaccine (PCV10) on a 6, 10, 18 weeks schedule in March 2015 as a second country in the South East Asia region after Pakistan.[2]

A significant advantage of Hib conjugate vaccine worldwide in resource-poor setting is the herd immunity, even at relatively low levels of vaccination coverage. The herd effect is the result of reduced nasopharyngeal colonization with the organism in vaccinated infants leading to transmission disruption. However, the greatest threat facing pneumococcal conjugate vaccine effectiveness is serotype replacement. The current vaccines provide serotype-specific, antibody-mediated protection against only a few of the 90+ capsule serotypes. Therefore, there has been a focus in recent years to rapidly advance technologies that will result in broader disease coverage and more affordable vaccines that can be used in developing countries. The next generation of pneumococcal vaccines have advanced to clinical trials.[3]

Global estimates for the year 2000 showed about 2.1 million severe infections and 299,000 child deaths from Hib, and 6.6 million severe infections and 600,000 child deaths from pneumococcus excluding the cases of opportunistic infection in children with HIV. The updated estimates on a country-by-country basis, for each year from 2000 to 2015 for the numbers of Hib and pneumococcal disease cases and deaths in children showed that both Hib and pneumococcus were responsible for far fewer cases of severe disease and deaths in children aged one to 59 months in 2015 compared to 2000. For Hib in 2015, there were approximately 29,500 child deaths, and for

1. Associate Professor and Head, Department of Pediatric Pulmonology, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital.
2. Professor & Head, Department of Microbiology, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital.

Correspondence to: Dr. Probir Kumar Sarkar, Associate Professor and Head, Department of Pediatric Pulmonology, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital. Cell: 01711225099, E-mail: drprobirdsh@gmail.com
Received: 13 August 2018; **Accepted**: 17 September 2018

pneumococcus an estimated 294,000 child deaths. These figures suggest declines of 90 percent and 51 percent, respectively, from the estimated deaths in the year 2000. [4] There was also a sharp reduction of deaths about 75 percent from Hib and pneumococcal diseases in children with HIV/AIDS from 95,000 in 2000 to about 23,000 in 2015.[1, 4]

In Bangladesh, the under five deaths due to acute respiratory infections was 63,294 in 2000, reduced subsequently to 27,800 in 2010 and 16,960 in 2016 suggests a significant fall in the under five mortality after the introduction of Hib and pneumococcal conjugate vaccine.[4,5]

The sharp declines in child mortality from Hib and pneumococcus were due not just to the introduction of vaccines but also to general factors that have reduced pneumonia and meningitis deaths from all causes, such as better hygiene and access to health care. However, there was evidence that vaccines were specifically responsible for a considerable reduction in mortality. The estimated average annual decline in child deaths from pneumococcus jumped from 3 percent during 2000-2010 to 8 percent after 2010 when many high-burden countries began widespread immunizations with PCV.[1,5]

Global data showed that during 2000-2015 PCV prevented a total of about 250,000 child deaths, mostly after 2010, while Hib vaccines prevented 1.2 million child deaths. These figures did not include the prevented cases of pneumococcal and Hib deaths among children who were HIV-infected.[1]

The new estimates will guide ongoing efforts to reduce the burdens of Hib and pneumococcal diseases, which together still kill approximately 900 children per day around the world. The estimates also suggest that about half of the pneumococcal child deaths in 2015 occurred in just four countries namely India, Nigeria, Democratic Republic of the Congo and Pakistan.[1,6]

Though there is remarkable impact of pneumococcal conjugate and Hib vaccines on controlling pneumonia, too many children are still not receiving the required vaccines today. It is anticipated that with accumulating evidence regarding vaccine effectiveness, herd immunity and surveillance assessments on the impact of pneumococcal conjugate vaccines on invasive disease, more countries will be motivated to include the vaccines into national immunization schedules. New opportunities are available to overcome the shortfalls of the current pneumococcal conjugate vaccines in the form of a new generation pneumococcal vaccines that are potentially more affordable and designed to maximize protection in developing countries.

References

1. Brain W, Katherine L O B, Adena G, Anwesha M, Li L, Yue C, Ivana L, et al. Burden of Streptococcus pneumoniae and Haemophilus influenzae type b disease in children in the era of conjugate vaccines: global, regional, and national estimates for 2000-15. *The Lancet* 2018; **6**: 744-56.
2. Shampa S, Hasan M, Lindsay K, Jennifer L, Belal H. Maksuda I ei al. Epidemiology and risk factors for pneumonia severity and mortality in Bangladeshi children <5 years of age before 10-valent pneumococcal conjugate vaccine introduction. *BMC Public Health* 2016; **16**: 1233.
3. UNICEF. Committing to Child Survival: A Promise Renewed: *Progress Report* 2015.
4. WHO and Maternal and Child Epidemiology Estimation Group (MCEE) Estimates 2018.
5. UNICEF Estimates of child causes of death, Acute Respiratory Infections 2018.
6. Diana C O, Allan W C. Vaccination for the control of childhood bacterial pneumonia- Haemophilus influenzae type b and pneumococcal vaccines. *Pneumonia 2013*; **14**(2): 2-15.1

DS (Child) H J 2018; 34(2) : 76-82

ORIGINAL ARTICLE

Immediate Outcome of Children Born with d-TGA: Experience in Dhaka Shishu Hospital Pediatric Cardiac Center

Abdul Jabbar[1], Nadia Binta Haoque[2] , Mohammad Abdullah Al Mamun[3], Manzoor Hussain[4], Rezoana Rima[5], Nowshika Sharmeen Echo[6]

Abstract

Background: *Transposition of the great arteries (TGA) is one of the commonest congenital cyanotic cardiac malformation. Without early recognition, diagnosis and treatment, a majority of infants and children with d-TGA die in their first month of life in developing countries.*

Objective: *This study was conducted to see immediate outcome of children born with d-TGA in the Pediatric Cardiac Center of Dhaka Shishu Hospital.*

Methods: *This prospective study was conducted in the Pediatric Cardiology center of Dhaka Shishu (Children) Hospital from July 2016 to July 2017. Child with d-TGA admitted for medical management and cardiac intervention during the study period were included. Data were collected from data collection sheet, OPD follow-up, hospital records and over telephone and analyzed by using SPSS version 17.*

Results: *Total 22 patients with d-TGA were admitted during the study period. Among them 68.2% were neonate and 31.8% were infant. Male were 54.5% and female were 45.5% with a male female ratio 1.2:1. Majority of children presented with cyanosis (40.9%) and heart failure (22.7%). Among them 63.6% were TGA with intact IVS & 36.4% were TGA with VSD. Ten out of 22 patients (45.5%) needs emergency Balloon Atrial Septostomy (BAS) intervention. Types of surgical intervention includes BAS followed by Arterial Switch Operation (ASO) in 36.4%, ASO without BAS in 18.1%, ASO with VSD Closure in 13.6% and Senning operation in 4.5%. Among surgical intervention (n-16), 9 patient underwent surgery in India, among them 6 survived and 3 expired and 7 patient underwent surgery in Bangladesh, among them 5 survived and 2 expired. Four out of 22 patients underwent no intervention and all of them died. Among 18 (81.8%) underwent surgical intervention, survival rate at 1 year of age is high (61.2%) and mortality rate is lower (38.8%). Also among the patients underwent BAS followed by ASO, survival rate is higher (75%) and mortality rate is lower (25%).*

Conclusion: *With the advent of early neonatal intervention and improved surgical techniques as well as post operative intensive care, the survival rate of d-TGA is satisfactory. To reduce the higher mortality among child with d-TGA early identification, early intervention, and referral to advanced cardiac center for early arterial switch operation is essential.*

Kew wards: *d-TGA, Balloon Atrial Septostomy, Arterial Switch Operation, Outcome.*

1. Registrar, Department of Pediatric Cardiology, Dhaka Shishu (Children) Hospital.
2. Resident Medical Officer, Department of Pediatric Cardiology, Dhaka Shishu (Children) Hospital.
3. Associate Professor, Division of Neonatal Cardiology, Department of Pediatric Cardiology, Bangladesh Institute of Child Health, Dhaka Shishu (Children) Hospital.
4. Head of Pediatric Medicine and Cardiology, Bangladesh Institute of Child Health, Dhaka Shishu (Children) Hospital
5. Associate Professor, Department of Pediatric Cardiology, Bangladesh Institute of Child Health, Dhaka Shishu (Children) Hospital.
6. Resident Medical Officer, Department of Pediatric Cardiology, Dhaka Shishu (Children) Hospital.

Address of correspondence: Dr. Abdul Jabbar, Registrar, Department of Pediatric Cardiology, Dhaka Shishu (Children) Hospital. Cell: 01720346665, E-mail: dr.zibon@yahoo.com

Received: 19 June 2018; **Accepted**: 07 August 2018

Introduction

d-TGA accounts for 5-7% of all congenital heart defects (CHD) with a prevalence of 0.2 per 1000 live births and male preponderance.[1] Sibling recurrence rates of 0.27 and 2%,respectively, have been noted in simple and complex forms associated with a functionalsingle ventricle or heterotaxy.[2] Causative mutations have been reported in the Nodal signaling pathway. Mutations in *ZIC3*, initially known to cause X-linked heterotaxy, have also been shown tocause sporadic and familial d-TGA. Abnormalities in other Nodal pathway genes, such as *CFC1* and *FoxH1*, have also been associated with isolated or syndromic d-TGA .[3-5] In this anomaly, the aorta arises from the morphological right ventricle (RV), and the pulmonary artery arises from the morphological left ventricle (LV) (i.e., there is ventriculoarterial discordance). Complete transposition of the great arteries is also known as d-TGA; the "d-" refers to the dextroposition of the bulboventricular loop (ie, the position of the RV, which is on the right side). The aorta also tends to be on the right and anterior, and the great arteries are parallel rather than crossing as they do in the normal heart. Because the systemic and pulmonary circulations run in parallel, there has to be a communication between the two, either with an atrial septal defect, a ventricular septal defect (VSD), or at the great arterial level (patent ductus arteriosus) to support life. These connections allow systemic blood to enter the pulmonary circulation for oxygenation and allow oxygenated blood from the pulmonary circuit to enter the systemic circulation. The most common associated lesions are VSD, which occurs in almost half of the cases, pulmonary outflow tract obstruction and, less commonly, coarctation of the aorta.[6]

The first successful arterial switch operation (ASO) was reported by Jatene et al[7] in 1975. Over the ensuing 3 decades, this procedure has come to be accepted as the preferred surgical treatment of the majority of children born with transposition of the great arteries (TGA) and forms of double outlet ventricle with subpulmonary ventricular septal defect, also known as the Taussig-Bing anomaly (TBA). Numerous publications have examined the role of the ASO in these conditions, and reported surgical outcomes have steadily improved with refinements in diagnosis, surgical technique, and perioperative management. A literature search lists over 180 articles published in the medical literature during the last 5 years focused on the subject of TGA and ASO.

The ASO benefits the patient by realigning the great vessel connections to provide ventriculoarterial concordance, an "anatomic" repair. This is in contrast to the atrial switch procedure (Mustard or Senning), which, although technically less challenging and providing "physiologic" correction, leaves the patient with ventriculoarterial discordance. The long-term detrimental effects of having a systemic right ventricle, particularly in patients with TGA with ventricular septal defect (VSD) are now well recognized and include dysrhythmia, tricuspid insufficiency and right ventricular failure.[8-10] In this regard, the ASO is clearly a superior option, affording the patient the benefit of a systemic left ventricle with normal circulatory pathways. In particular, certain anatomic substrates have been associated with increased operative risk, including patients with complex coronary origin and branching patterns, patients with associated aortic arch anomalies and left ventricular outflow tract (LVOT) obstruction.[11-14]

In developing nations with limited infrastructure, human, and material resources, pediatric cardiac intensive care is yet to take roots as a distinctive discipline. The services are provided by a multidisciplinary team that includes pediatric cardiologists, pediatric cardiac surgeons, intensivists, critical care nurses, respiratory therapists, and other support personnel.[15] With the scope of complete correction, more and more children are undergoing cardiac interventions and surgical treatment for CHD, so there is increasing demand for dedicated personnel for the specialized intensive care of these critically ill children.[16]

The long-term outcomes of patients with d-TGA have improved dramatically in the current ASO era with advances in prenatal diagnosis, BAS, PGE, surgical technique and postoperative management. The trend toward early cardiac intervention and primary surgical repair during early infancy or newborn period is now well established.[17]Without early recognition, diagnosis and treatment, a majority of infants and children with CHD die in their first month of life in developing countries.[18]

Bangladesh is facing a multitude of health problems and congenital heart disease (CHD) is one of them. Hussain et al[19] during early nineties found only 8.3% CHD at neonatal period admitted in Dhaka Shishu Hospital. During January 1998 to December 1999

11.9% CHD were diagnosed during neonatal period and during January 2008 to December 2009 number increased to 27.5% .[20]As without early recognition, diagnosis and treatment, a majority of infants and children with d-TGA die in their first month of life in developing countries this study was conducted to seeimmediate outcome of children born with d-TGA in Pediatric Cardiac Center of Dhaka Shishu Hospital.

Materials and Methods

This prospective study was conducted in the Pediatric Cardiology Center of Dhaka Shishu (Children) Hospital from July 2016 to July 2017. Children with d-TGA admitted for medical management and neonatal cardiac interventions (BAS) were included. Primary cardiac diagnosis was confirmed in all cases by transthoracic 2D and color doppler echocardiogram. Data were collected from data collection sheet, hospital records, OPD follow up at 1 year of age, over telephone and analyzed by using SPSS version 17.

Results

Total 22 child with d-TGA were admitted during the study period. Male were 54.5% and female were 45.5 % with a male female ratio 1.2:1. Among them 15 (68.2%) were neonate and 7(31.8%) were diagnosed after neonatal period (Table-I).

Table-I
Demography of study population (n=22)

Baseline characteristics		Frequency	%
Sex	Male	12	54.5
	Female	10	45.5
Age	Neonate	15	68.2
	Infant	06	31.8

Majority of children presented with cyanosis 9(40.9%), heart failure 5 (22.7.%), respiratory tract infection 5(22.7%) and cardiac murmur 3 (13.6%) (Fig.-1).

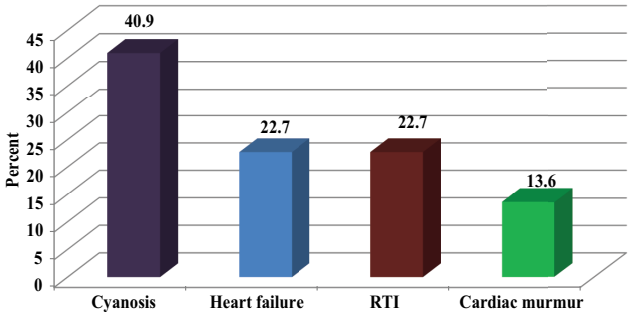

Fig 1 *Distribution of initial clinical presentation of d-TGA among admitted children*

Among the admitted patients 14 (63.6%) were TGA with intact IVS and 8 (36.4%) were TGA with VSD (Table-II).

Table II
Distribution of types of d-TGA (n=22)

Varieties	Number (%)
D-TGA, Intact IVS, PFO	7 (31.8)
D-TGA, Intact IVS,PFO, PDA	5(22.8)
D-TGA, Intact IVS, Large ASD	2 (9.10)
D-TGA, VSD, PFO	3 (13.6)
D-TGA, VSD,PFO, PDA	2 (9.10)
D-TGA,VSD, PS	3(13.6)

During the study period BAS was done in 10 cases, among them ASO done in 8 cases, 4 patient underwent ASO without BAS, 3 patient underwent ASO+VSD closure,1 patient underwent Senning procedure and 4 patient underwent no intervention (Table-III). All the cases of BAS is performed at our centre. Among surgical intervention (n-16), 9 patient underwent surgery in India, among them 6 survived and 3 expired and 7 patient underwent surgery at National Heart Foundation Hospital and Research Institute (NHFH&RI), among them 5 survived and 2 expired.

Table-III
Outcome of cardiac intervention & surgery among the d-TGA patient (n=22)

Interventions/Surgery	Number (%)	Immediate outcome	
		Survived	Expired
BAS	10(45.5%)	08(80.0%)	02(20.0%)
BAS followed by ASO	08(36.4%)	06(75.0%)	02(25.0%)
ASO without BAS	04(18.1%)	02(50.0%)	02(50.0%)
ASO + VSD Closure	03(13.6%)	02(66.7%)	01(33.3%)
Senning Operation	01(4.5%)	01(100%)	0 (0%)
No intervention/Surgery	04(18.1%)	0 (0%)	04(100%)

BAS: Balloon Atrial Septostomy, AS0: Arterial Switch Operation

Among the d-TGA patients 11 (50.0%) were survived and 11(50.0%) were found dead at 1 year follow up (Fig.-3).

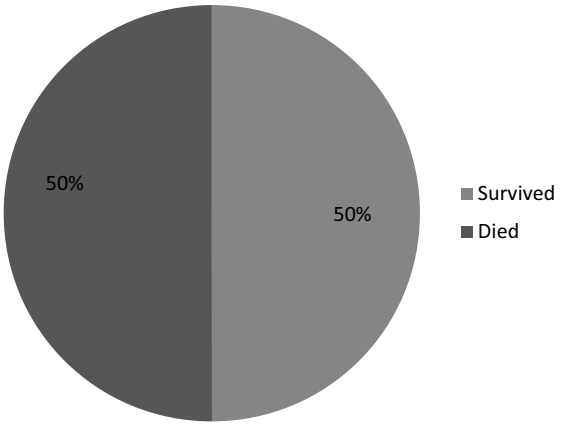

Fig 3 *Outcome of patients with d-TGA (n=22)*

Out of 22 admitted patients with d-TGA, 18 (81.8%) underwent surgical intervention. Among them survival rate is high (61.2%) and mortality rate is lower (38.8 %) (Fig.-4).

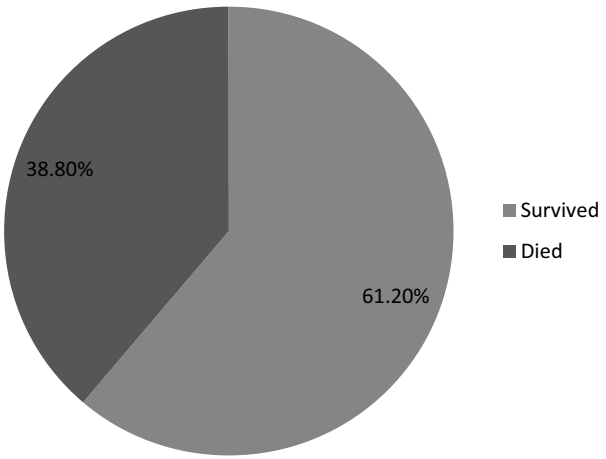

Fig 4 *Outcome of patients with d-TGA underwent surgical intervention (n=18)*

Out of 22 admitted patients with d-TGA, 10 (45.5%) underwent BAS, among them 8 underwent ASO. Survival rate is higher among patient underwent BAS followed by ASO. Out of 8 patients, 6 (75%) patient survived and 2 (25%) patient expired (Fig.-5).

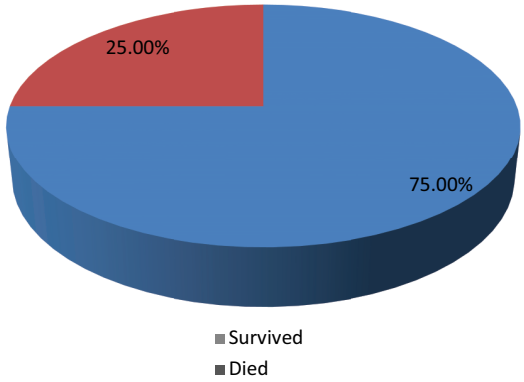

Fig 5 *Outcome of patients with d-TGA underwent BAS followed by ASO (n=08)*

Total 15 patients underwent ASO, among them BAS followed by ASO done in 8 patient and 7 patient underwent ASO without BAS. Although survival rate is higher among BAS followed by ASO group, χ^2 test is done between these two group for outcome but P-value is found to be statistically insignificant (Table-IV).

Table-IV

Child with d-TGA underwent ASO with or without BAS & there outcome (n=15)

Group	Surgical intervention (n-15)	Survived	Died	P*
BAS followed by ASO	8 (53.33)	6(75)	2(25)	0.03
ASO without BAS	7(46.67)	4(57.2)	3(42.8)	

*χ^2 test

Co morbid conditions like metabolic acidosis, heart failure, pneumonia, shock and sepsis also contributed in mortality (Fig.-6)

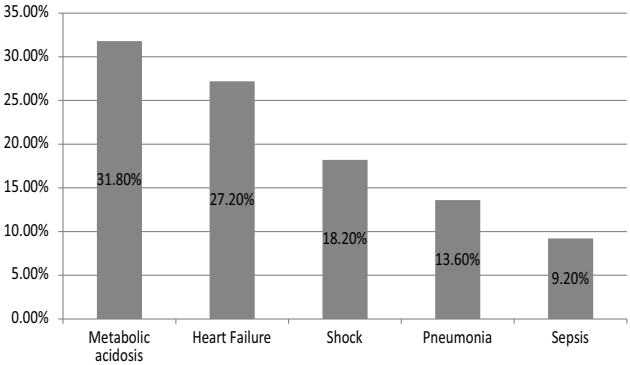

Fig 6 *Co-morbid conditions among child with d-TGA*

Discussion

d-TGA is one of the commonest cyanotic heart disease in neonate where early identification, early intervention and surgical correction is necessary. In the developed world Critical Congenital Heart Disease (CCHD) can be treated with early surgery or transcatheter interventions. In the current era, congenital heart surgery allows for repair or palliation of nearly all types of congenital heart malformations. Congenital heart surgery, together with transcatheter interventions, has resulted in a marked improvement in survival for those with CCHD. Intervention is typically performed in the first weeks of life to optimize hemodynamics and prevent end-organ injury associated with delayed diagnosis. With the advent of prostaglandin therapy for ductus arteriosus-dependent lesions, many previously lethal congenital heart conditions that present with severe hypoxemia, shock, and acidosis in the newborn period are now survivable and can be palliated.[21]

This prospective study was conducted among children with d-TGA admitted during the study period. Primary cardiac diagnosis was confirmed in all cases by transthoracic 2D echocardiogram. Total 22 patients with d-TGA were admitted during the study period. Among them 68.2% were neonate and 31.8% were infant. Male were 54.5% and female were 45.5% with a male female ratio 1.2:1. Samanek et al[1] also showed male predominance in their study. Majority of children presented with cyanosis (40.9%) and heart failure (22.7%). Among the admitted children 63.6% were TGA, Intact IVS & 36.4% were TGA with VSD which is similar to Hasan et al[22] study reported 50% had TGA with Intact IVS, 34.6% TGA with VSD, 11.5% Taussig Bing anomaly and 3.8% TGA with complex cardiac anomaly. Dibardinoet al[23] showed primary cardiac diagnoses included TGA with intact ventricular septum (TGA/IVS, n = 79, 63%), TGA with ventricular septal defect (TGA/VSD, n = 37, 30%), and Taussig Bing Anomaly (TBA, n = 9, 7%). Brown et al[24] reported the patient population included TGA with intact ventricular septum (58.7%, 118 of 201), with ventricular septal defect (31.3%, 61 of 201), and Taussig-Bing anomaly (10.0%, 22 of 201).

In these study 10 out of 22 patients (45.5%) needs emergency BAS intervention. BAS was not performed in patients with an obviously nonrestrictive atrial septal defect. BAS is an important component of the preoperative management strategy. The presence of an unrestrictive ASD often allows patients to be weaned from PGE1, extubated, and fed orally before surgery. Dibardino et al[23] study showed preoperative BAS was performed before operative intervention in 109 cases (87%) to optimize the opportunity for atrial level mixing.

Arterial switch during the neonatal period has become the treatment of choice for d-TGA in most hospitals because of the excellent surgical outcome and the low perioperative mortality reported by experienced surgical teams.[25,26] In these studytypes of surgical intervention includes BAS followed by ASO (36.4%), ASO without BAS (18.1%), ASO with VSD closure (13.6%) and Senning operation (4.5%). The surgical management of transposition of the great arteries and intact ventricular septum (TGA/IVS) beyond 2 to 3 weeks of age is controversial. Concern that regression of the left ventricular (LV) myocardialmass will render the left ventricle incapable of coping with the acutely increased work of systemic perfusionhas been considered a contraindication to a primary ASO.[27] Options available after LV regression are eitheratrial switch procedures like Senning or Mustard operation or left ventricular training followed by arterials witch. The results of ASO had been encouraging ascompared to the atrial switch operations. Worldwide it was accepted that ASO is superior to Senning/Mustard operations.[28] Among the admitted patients with d-TGA 11(50.0%) were survived, 11(50.0%) died. Among the patients underwent surgical intervention 18(81.8%), survival rate is high (61.2%) and mortality rate is lower (38.8%). Four out of 22 patients underwent no intervention due to financial problem and all of them died. Co morbid condition like metabolic acidosis, heart failure, shock, pneumonia and sepsis also contributed in mortality. Rodríguez et al[29] reportedthe overall operative mortality rate was 11.6% over the 25-year period. Brown et al[24] report overall, early mortality was 9.5% (19 of 201) and there were five late deaths (2.7%). One-month, 1-year, and 5-year actual survival rates were 90.4%, 87.9%, and 87.9%, respectively. The survival rate of children with d-TGA and an intact interventricular septum improved dramatically after balloon atrial septostomy. Total 15 patients underwent ASO, among them BAS followed by ASO done in 8 patient and 7 patient underwent ASO without BAS. Survival rate is higher (75%) among patients underwent BAS followed by ASO in comparison to ASO without BAS group. This finding is statistically significant ($p<0.05$).

Conclusion

With early surgical intervention and cardiac the survival rate of d-TGA is satisfactory. To reduced the higher mortality among child with D-TGA early identification, early intervention and referral to advanced cardiac center for early arterial switch operation is essential. Further improvements may be achieved by reinforcing prenatal diagnosis and by establishing strategies to manage baby born with d-TGA.

References

1. Samanek M. Congenital heart malformations: prevalence, severity, survival, and quality of life. *Cardiol Young* 2000; **10**: 179-85.
2. Digilio MC, Casey B, Toscano A. Complete transposition of the great arteries: patterns of congenital heart disease in familial precurrence. *Circulation* 2001; **104**(23): 2809-14.
3. Unolt M, Putotto C, Silvestri LM. Transposition of great arteries: new insights into the pathogenesis. *Front Pediatr* 2013; **1**: 11.
4. De Luca A, Sarkozy A, Consoli F. Familial transposition of the great arteries caused by multiple mutations in laterality genes. *Heart* 2010; **96**: 673-77.
5. D'Alessandro LC, Latney BC, Paluru PC, Goldmuntz E. The phenotypic spectrum of ZIC3 mutations includes isolated d-transposition of the great arteries and double outlet right ventricle. *Am J Med Genet A* 2013; **161**A(4): 792-802.
6. Warnes CA. Transposition of the great arteries. *Circulation* 2006; **114**(24): 2699-09.
7. Jatene AD, Fontes VF, Paulista PP. Anatomic correction of transposition of the great vessels. A preliminary report. *Ar Qbras Cardiol* 1975; **28**: 461-64.
8. Gelatt M, Hamilton RM, McCrindle BW. Arrhythmia and mortality after the Mustard procedure: A 30-year single center experience. *J Am Coll Cardiol* 1997; **29**: 194-201.
9. Kirjavainen M, Happonen JM, Louhimo I. Late result of Senning operation. *J ThoracCardiovasc Surg* 1999; **117**: 488-95.
10. Turina MI, Sibenmann R, von Segesser L, Schonbeck M, et al. Late functional deterioration after atrial correction for transposition of the great arteries. *Circulation* 1989; **80**: 162-67.
11. Daebritz SH, Nollert G, Sachweh JS. Anatomic risk factors for mortality and cardiac morbidity after arterial switch operation. *Ann Thorac Surg* 2000; **69**: 1880-86.
12. Planche C, Bruniaux J, Lacour-Gayet F. Switch operation for transposition of the great arteries in neonates. *J Thorac Cardiovasc Surg* 1988; **96**: 354-63.
13. Scheule AM, Zarakowski D, Blume ED. Arterial switch operation for a single coronary artery. *J Thorac Cardiovasc Surg* 2002; **123**: 1164-72.
14. Day RW, Laks H, Drinkwater DC. The influence of coronary anatomy on the arterial switch operation in neonates. *J Thorac Cardiovasc Surg* 1992; **104**: 706-12.
15. Chang AC. Starting a pediatric cardiac intensive care program: Essential elements for sustained success. *Prog Pediatr Cardiol* 2003; **18**: 123-30.
16. Stromberg D. Pediatric cardiac intensivists: Are enough being trained? *Pediatr Crit Care Med* 2004; **5**: 391-92.
17. Fuchs RJ, Berenholtz SM, Dorman T. Do intensivists in ICU improve outcome? B*est Pract Res Clin Anaesthesiol* 2005; **19**: 125-35.
18. United Nations 2000. Millennium development goals: UN Millennium Declaration. http://www.un.org/millenniumgoals/ (accessed March 14, 2015).
19. Hussain M, Tahura S, Sayeed MA, Rahman MM, Rahman MM, Kar SK. Past and present pattern of congenital heart disease at Dhaka Shishu Hospital: A situation analysis. *Bangladesh Journal of Child Health* 2010; **34**: 51-55.
20. Hussain M, Tahura S, Hussain MZ, Fatema NN, Razzaque SKA. Pattern of congenital heart disease in bangladesh: A multi-center study. *DS (Child) H J* 2011; **27**: 5-11.
21. Rashkind WJ, Gibson J Jr. Catheter interventions in congenital heart defects as alternatives to open surgery. *Lakartidningen* 2005; **102** :2060-63.
22. Hasan MNA, Sharifuzzaman M. Surgical Management of TGA at National Heart Foundation Hospital: Nouveau Espoir. *Paed. Heart J of Bangladesh (PHJB)* 2017; **2**(1): 16-25.
23. Dibardino DJ, Allison AE, Vaughn WK, McKenzie ED, Charles D. Current expectations for newborns undergoing the arterial switch operation. *Ann Surg* 2004; **239**(5): 588-98.

24. Brown JW, Park HJ, Turrentine MW. Arterial switch operation: Factors impacting survival in the current era. *Ann Thorac Surg* 2001; **71**(6): 1978-84.

25. Losay J, Touchot A, Serraf A, Litvinova A, Lambert V, Piot JD, et al. Late outcome after arterial switch operation for transposition of the great arteries. *Circulation* 2001; **104**(12): 121-26.

26. Oda S, Nakano T, Sugiura J, Fusazaki N, Ishikawa S, Kado H. Twenty-eight years experience of arterial switch operation for transposition of great arteries in a single institution. *Eur J Cardiothorac Surg* 2012; **42**:674-9.

27. Bisoi AK, Chauhan S, Khanzode SD, Hote MP, Juneja R. D-Transposition of great vessels with intact ventricular septum presenting at 3 – 8 weeks: Should all go for rapid two stage arterial switch or primary arterial switch? *Ind J Thorac Cardiovas Surg* 2006; **22**: 5-9.

28. Kang N, de Leval MR, Elliott M, Tsang V, Kocyildirim E, Sehic I, et al. Extending the boundaries of the primary arterial switch operation in patients with transposition of the great arteries and intact ventricular septum. *Circulation* 2004; **110**: 123-27.

29. Rodríguez MJ, Letrán LC, Vazquianez MR, De Soto JS, Hosseinpour R et al. Mid-term morbidity and mortality of patients after arterial switch operation in infancy for transposition of the great arteries. *Rev Esp Cardiol* 2014; **67**(3): 181-88.

DS (Child) H J 2018; 34(1) : 83-87

ORIGINAL ARTICLE

Iron Deficiency Anaemia of Children Attending at Out Patient Department of Dhaka Shishu (Children) Hospital

Mahbubur Rahman[1], Rabi Biswas[2], Manzoor Hussain[3]

Abstract

Background: *Iron deficiency anemia, a major public health problem in our country like other developing countries. It is associated with poor growth and developmental outcome in children.*

Objective: *This study was done to find out the extent and severity of iron deficiency anaemia with view to overcome the problem and to improve nutritional status of children.*

Methods: *It is a cross sectional study, conducted from January 2015 to June 2015. Children attended at outpatient department of Dhaka Shishu Hospital (DSH) were included. Written consent along with prior approval of ethical review committee and preformed questionnaire was used to take details of patient. Blood was collected, centrifuged, and was analyzed for complete blood count with peripheral blood film, serum iron and serum ferritin concentration.*

Results: *Out of total 74 children, 59 children (68%) were found to be anaemic and 47 of children (63%) were iron deficient.*

Conclusion: *Iron deficiency anaemia is a major public health problem. Majority of the patient (68%) was found to anemic and iron deficient (63%). This issue should be taken into consideration to over come the problem.*

Key words: *Iron deficiency, Iron deficiency anaemia.*

Introduction

Iron deficiency (ID) and iron-deficiency anaemia (IDA) continue to be of worldwide concern. Among children in the developing world, iron is the most common single-nutrient deficiency.[1] In industrialized nations, despite a demonstrable decline in prevalence[2], IDA remains a common cause of anaemia in young children. However, even more important than anaemia itself is the indication that the more common ID without anaemia may also adversely affects long-term neurodevelopment and behavior and that some of these effects may be irreversible.[3-4] A haemoglobin (Hb) concentration 2 SDs below the mean Hb concentration for a normal population of the same gender and age range, as defined by the World Health Organization, the United Nations Children's Fund,

1. Assistant Professor, Department of Pediatric Hepatology, Gastroenterology & Nutrition, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital.
2. Assistant Professor, Department of Pediatric Endocrinology & Metabolic Disorder, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital.
3. Professor and Head, Department of Pediatric Medicine and Pediatric Cardiology, Bangladesh Institute of Child Health (BICH) and Dhaka Shishu (Children) Hospital, Dhaka.

Correspondence to: Dr. Md. Mahbubur Rahman**,** Assistant Professor, Department of Pediatric Hepatology, Gastroenterology & Nutrition, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital. Cell: 01711704701, Email: mahbub.rahman1028 @gmail.com

Received: 13 January 2018; **Accepted**: 11 April 2018

and United Nations Universit.[5] On the basis of the 1999-2002 US National Health and Nutrition Examination Survey, anaemia is defined as a Hb concentration of less than 11.0 g/dL for both male and female children aged 12 through 35 months. For certain populations (i.e., people living at high altitudes), adjustment of these values may be necessary.[6-7]

The term iron deficiency refers to a state in which the body iron stores have been depleted, it implies neither the degree of depletion not the presence of anaemia. Iron deficiency anaemia refers to a haemoglobin state resulting from deficiency. It's occurrence implies iron stores are severely depleted. Thus an individual may be iron deficient without manifesting iron deficiency anaemia.[8] Iron deficiency is the result of an iron imbalance i.e., the amount absorbed by the intestine may not be satisfactory to meet body requirements. However, if the supply of haemopoietic nutrients is inadequate the bone is dysfunctional, the level of haemoglobin will be subnormal and a state of anaemia will set in.

Iron requirements depends on age, sex, race, pregnancy, lactation and altitude. Because of the contributing factors for the development of anaemia in children information of its prevalence, distribution and aetiology are important for adopting appropriate measure for its prevention and control. During infancy the blood volume becomes double within 5-6 months, but the iron store in the reticulo-endothelial system is not adequate enough to cope with the increased demand. Moreover, during this period the child is fed absolutely on milk (either cow's milk or mother's milk) which is deficient in iron. If mixed feed is not started earlier as milk powder supplemented with iron is not given then anaemia develops.[9]

Serum feritin (SF) is a sensitive parameter for the assessment of iron stores in healthy subjects,[10-12] 1μg/L of SF corresponds to 8 mg of available storage iron.[13-15] Measurement of SF concentration is widely used in clinical practice and readily available. Cook et al[16] selected an SF concentration below 12 μg/L as diagnostic for ID after a comprehensive population survey in the United States. Thus, a cutoff value of 10 μg/L has been widely used for adults and denotes depletion of iron stores. In children, a cutoff value of 10 μg/L has suggested.[17] Combining SF concentration with a determination of CRP is currently more readily available to assess iron stores and is a reliable screening test as long as the CRP level is not elevated.[18]

The SF falls with iron deficiency but rises with inflammation, although the SF level that signifies iron deficiency may be altered by chronic inflammation. It is still a useful measure of iron status.[19] A decline in serum ferritin is very important indication of developing iron deficiency adults, values less than 12 ng/ml are considered to indicate depletion of iron store. The cause of anaemia can be reasonably attributed to iron deficiency only when at least two iron parameters fall within the iron deficient range. The study was done to find out in prevalence of anemia and iron deficiency.

Materials and Methods

A cross sectional study was carried out from January 2015 to June 2015 in out patient department of Dhaka Shishu Hospital. Children attended at OPD of Dhaka Shishu Hospital were included in this study. Children with hamatological disorder like thalassaemia, sickle cell anaemia and patient with severe infection were excluded from the study. A structured questionnaire was developed to obtained relevant information, socio-economic history and dietary history. Study participants were selected randomly. Written consent was taken and prior approval from the ethical review committee was obtained. About 7ml blood was collected aseptically, one ml of blood was transferred to a clean glass vial containing EDTA for hematological examination. The rest of the blood was transferred into centrifuge tube, allows to stand for 2 hours at room temperature and the centrifugation was done at the rate of 3000 r.p.m for 10 minutes, serum sample was separated, frozen, and stored at 20pc for further analysis. The investigation which were done include complete blood count (CBC) with peripheral blood film (PBF), serum ferritin and serum iron.

Results

Table- I
Distribution of age, number and percentage of children

Age in (months)	Number	Percentage
5-24	34	45.9
25-60	25	33.8
61 +	15	20.3
Total	74	100

Table I shows total number of children in the study. Among them in the age group 5-24 month number of children was 34 (45.9%) in the age group 25-60 month number of children was 25(33.8%) and 61 month and above the cause was 15(20.3%).

Table II
Number and percentage distribution of gender of study participants by age group

Age of the children (months)	Male		Female	
	n	%	n	%
5-24	20	50.0	14	41.2
25-60	9	22.0	16	47.1
61 +	11	27.0	4	11.8
	40	100	34	100

Table II shows distribution of age, number and percentage of study participants. The number of male children in the age group 5-24 months were 20 (50%). Among the age group 25-60 months the number were 9 (22%) and in the age group 61 (+) the number was 11 (27%). Total female children in the study were 34 (45.95%). The number of children in the age group 5-24 months were 14 (41.2%) of age group 25-60 the number were 16 (47.1%) and in the age group 61 month (+) cases was 4 (11.8%).

Table III
Distribution of anaemia of the study participant by Hb level (gm/dL)

Hb level	Number	Percentage
< 11 gm/dL	59	68
≥11 gm/dL	15	32
Total	74	100

Table III shows distribution of number and percentage, anemia of study participant. Among the study participant 59 (68%) had haemoglobin level <11gm/dL and 15 children (32%) had hemoglobin level ≥12gm/dL.

Table IV
Peripheral blood film criteria of study participant

Blood film criteria	Number	Percent
Normocytic normochromic	15	20.3
Microcytic hypochromic	49	66.2
Dimorphic RBC	10	13.5
Total	74	100

Table IV shows 15 children had normocytic normochromic blood picture and 49 of the children (66.2%) had microcytic blood film and 10 (13.5%) of children had dimorphic blood film.

Table V
Serum ferritin level of study participant

Ferritin level	Number	Percentage
<12 ng/mL	47	63
>12 ng/mL	27	37
Total	74	100

Table V shows serum Ferritin level of the study participants. 47 children (63%) of the children had serum iron concentration <40µg/dL and 27 children (37%) had serum concentration >40 µg/dL.

Table VI
Serum iron level of study participant (ìg/dL)

Iron level	Number	Percentage (%)
< 40/ìg/dL	47	63
> 40/ìg/dL	27	37
Total	74	100

Table VI shows serum iron level of the study participants. 47 children (63%) of the children had serum iron concentration <40µg/dL and 27 children (37%) had serum concentration >40µg/dL.

Discussion

It is a prospective study which was done in out patient department of DSH to get information regarding incidence of iron deficiency anaemia in children. Iron deficiency is highly prevalent throughout the world and in public health term, iron deficiency is by far the most common cause of nutritional anaemia.[21] Haemoglobin level below the acceptable normal value for the definition of anaemia may not be necessarily to indicate iron deficiency. Since other factors such as folate deficiency, protein calorie malnutrition, chronic infection and inflammatory disease may also cause anaemia.[22]

The present study was done to diagnose and investigate anaemia and iron deficiency anaemia of children 5 month to 144 months. The children who were included in this study may not reflect the over all population from which they were drawn because

only the children reporting to the hospital for treatment were included in this study. So one should be cautious in interpretating the data, as not all children in this community were included.

According to WHO, children 6 months to 6 years with haemoglobin level below 11 g/dL considered to be anaemic. In the present study we found that 68% of the children were anaemic. Also by WHO criteria haemoglobin level of the age group 6-12 years below 12 gm/dL considered to below normal.[23]

According to Nutrition survey of Rural Bangladesh 1975-76, the children of the age group 0-4 years 82% of the children were anaemic and among age group 5-14 years, 74% of boys,75% of girls were anaemic.[24]

According to Nutrition survey of Rural Bangladesh 1981-82 the prevalence of anaemia in the age group 0-4 years and 5-40 years were 73% and 74% respectively.[25] Ayesha Molla et al. conduct a survey to estimate the prevalence of anaemia in urban slums and showed that 70% of the children of the age group 6-60 months were anaemic.[26]

According to 1988 national Nutrition Survey of Pakistan 65% of the children aged 7-60 months were anaemic. In India 52% of the pre-school children were found to be anaemic.[27] The mean haemoglobin level of 1975-76 survey showed it was 9.7 gm/dL 100 mL by the age group 0-4 years. We attempted in the present study to investigate iron nutrition status in relation to anaemia. In recent years several bio-chemical parameters are used for screening iron deficiency.

We measured serum iron best, known parameter to identify uncomplicated iron deficiency. In population where chronic infections may contribute to anaemia serum ferritin, an indicator of stored iron, would be the best parameter to diagnose the iron deficiency anaemia, as serum ferritin level falls only in true iron deficiency but not in chronic infection.[28]

At least two abnormal parameters of iron status have now been suggested to assess the iron deficiency anaemia in a population because anaemia may also be caused by other factors like inflammatory disease.

Using the acceptable cut off point for serum iron values (<40.0 μg/dL) about 63% of the children were found to have iron deficiency in this study.[29] When blood film criteria was used as an indicator 66% of the children were found to be iron deficient.

A study in neighboring Pakistan by Ayesha Molla et al. on the age group 6-60 months in an Urban slums of Karachi showed that 61% of the children children were classified as microcytic.[30] A study by Jalil and Khan showed that 83% of the children between 1-5 years of age were found to suffer from iron deficiency anaemia.[31] The above two study of iron deficiency anaemia more or less correlates with our study by their prevalence.

Conclusion

Iron deficiency anaemia is a major public health problem. Majority of the patient (68%) was found to anemic and iron deficient (63%). This issue should be taken into consideration to over come the problem.

References

1. United Nations Administrative Committee on Coordination Sub-Committee on Nutrition and International Food Policy research Institute. Fourth Report of the World Nutrition Situation. Geneva, Switerland: United Nations Administrative Committee on Coordination/Sub-Committee on Nutrition; 2000.
2. Sherry B; Mei Z, Yip R. Continuation of the decline in prevalence of anemia in low-income infants and children in five status. *Pediatrics* 2001; **107**(4): 677-82.
3. Lozoff B, Jimenez E, Smith JB. Double burden of iron deficiency in infancy and low socioeconomic status: a longitudinal analysis of cognitive test scores to age 19 years. *Arch Pediatr Adolesc Med* 2006; **160**(11): 1108-13.
4. Bruner AB, Joffe A, Duggan AK, Casella JF, Brandt J. Randomized study of cognitive effects of iron supplementation in non-anemic iron deficient adolescent girls. *Lancet* 1996; **348**(99033): 992-96.
5. American Academy of Pediatrics, Committee on Nutrition, Iron fortification of infant formulas. Pediatrics 1999; **104** (1pt 1): 119-23.
6. World Health Organization. Iron Deficiency Anemic: Assessment, Prevention, and Control-A Guide for Program Managers, Geneva, Switzerland: World Health Organization; 2001,Who/NHD/01.3.
7. Centers for Disease Control and Prevention National Center for Health Statistics. National Health and Nutrition Examination Survey. Available at: www.edc.gov/nchs/ nhanes.htm. Accessed September 29: 2008.
8. Hereberg S. Iron and folate deficiency anaemias children in the tropics. Review of the Internatioinal

Children's Center 1990: No.186: 4-40.

9. Khan MR, Nahar N, Rahman A, Sarker NI. Anaemia in childhood and infancy. *Journal of the Diabetic Association of Bnagladesh* 1977: **12**(2,3): 10.
10. Jacobs A, Miller F, Worwood M, Beamish MR, Wardrop CA. Ferritin in the serum of normal subjects and patients with iron deficiency and iron overload. *Br Med J* 1972; **4**(5834): 206–208.
11. Walters GO, Miller FM, Worwood M. Serum ferritin concentration and iron stores in normal subjects. *J Clin Pathol* 1973; **26**(10): 770-72.
12. Cook JD, Lipschitz DA, Miles LE, Finch CA. Serum ferritin as a measure of iron stores in normal subjects. *Am J Clin Nutr* 1974; **27**(7): 681-87.
13. Jacobs A, Miller F, Worwood M, Beamish MR, Wardrop CA. Ferritin in the serum of normal subjects and patients with iron deficiency and iron overload. *Br Med J* 1972; **4**(5834): 206-08.
14. Birgegård G, Högman C, Killander A, Levander H, Simonsson B, Wide L. Serum ferritin and erythrocyte 2,3-DPG during quantitated phlebotomy and iron treatment. *Scand J Haematol* 1977; **19**(4): 27.
15. Jacob RA, Sandstead HH, Klevay LM, Johnson LK. *U*tility of serum ferritin as a measure of iron deficiency in normal males undergoing repetitive phlebotomy. *Blood* 1980; **56**(5): 786-91.
16. Cook JD, Lipschitz DA, Miles LE, Finch CA. Serum ferritin as a measure of iron stores in normal subjects. *Am J Clin Nutr* 1974; **27**(7): 681-87.
17. Dallman PR, Siimes MA, Stekel A. Iron deficiency in infancy and childhood. *Am J Clin Nutr* 1980; **33**(1): 86-118.
18. Brugnara C, Zurakowski D, DiCanzio J, Boyd T, Plant O. Reticulocyte hemoglobin content to diagnose iron deficiency in children, *JAMA* 1999; **281**(23): 2225-30.
19. Birgegård G, Högman C, Killander A, Levander H, Simonsson B, Wide L. Serum ferritin and erythrocyte 2,3-DPG during quantitated phlebotomy and iron treatment. *Scand J Haematol* 1977;**19**(4): 327.
20. Dallman PR, Siimes MA, Stekel A et al. Iron deficiency in infancy and childhood. *Am J Clin Nutr* 1980; **33**(1): 86-18.
21. Hereberg S. Iron and folate deficiency anaemias children in the tropics. Review of the International Children's Center:1990: No.186: 4-40.
22. WHO (1972) Nutritional anaemia T. Echa.Rep. Ser.No.503 WHO, Geneva.
23. Jelliffe DR, Jelliffe EFP. Community Nutritional Assessment, 1989:**501**: Oxford University pren, October, U.K.
24. Nutrition survey of Rural Bangladesh, 1975-76. Institute of Nutrition and Food Science (INFS) University of Dhaka, Bangladesh, December 1977.
25. Nutition Survey off Rural Bangladesh 81-82. Institute of nutrition and food science Unbiversity of Dhaka, Bangladesh, December 1983.
26. Zeorge Buchanam and Riehard G. Sheehan. Malabsorption and defective utilization of iron in three sibling. *The Journal of Pediatrics* 1988; **8**: 723-28.
27. National Nutrition Survey 1985-**87**. Nutrition Division, National Institute of Health Government of Pakistan 1988:35.
28. Cart5 wright, GE, Lee GR. Annotation the anaemia of chronic disorders: 17 Brit. S. Haemato, 1979: **21**:147-55.
29. Cook JD, Finch CA. Assessing iron status of a population. *Am J Clin Nutr* 1979; **32**: 2115-2119.
30. Dallman PR, Yip R, Johnson C. Prevalence causes of anaemia in the united states 1976 to 1980. *Am J Clis Nutr* 1984; **39**: 437-45.
31. Jalil F, Khan AM. Nutritional anaemia Classification and effect of a therapeutic trial for proposed fortification programme. Islamabad, Planning and Development Division, Nutrition Cell, Government of Pakistan,1986-1988:15.

DS (Child) H J 2018; 34(1) : 88-93

ORIGINAL ARTICLE

Changes in Electrolyte in Different Stages of Perinatal Asphyxia: A Hospital Based Study

Sheikh Farjana Sonia[1], Soma Halder[2], Ruma parvin[1], Azmeri Sultana[3], Shyla Rahman[4], Farhana Rahat[1]

Abstract

Background: *Perinatal asphyxia is the most common cause of neonatal morbidity and mortality in worldwide. Electrolyte abnormalities are common in the immediate post asphyxiated period and influence the neonatal outcome effectively. So early recognition and prompt treatment of electrolyte changes is essential for optimal management and satisfactory long term outcome.*

Objective: *The aim of this study was to determine the electrolyte (sodium, potassium, calcium) changes in asphyxiated newborns of different severity in the early neonatal period and to find out the correlation of levels of sodium, potassium and calcium with the severity of perinatal asphyxia.*

Methods: *This cross sectional study was conducted in special care neonatal ward of Dr. M. R. Khan Shishu Hospital & Institute of Child Health, Mirpur-2, Dhaka over a period of six months from September 2017 to February 2018. A total of 120 neonates with Perinatal asphyxia HIE stage I, HIE stage II and HIE stage III presented within 3 days of life were included. Demographic profile, clinical presentations were taken by a structured questionnaire and serum electrolytes were measured and values were compared with the different severity of asphyxia. Data were analyzewd by using SPSS version 16.*

Results: *Among the 120 asphyxiated neonates 35% had hyponatremia, 30% had hyperkalaemia and 33.3% presented with hypocalcaemia. The mean values of sodium for HIE stage I, II and III were 135.62±5.51mmol/l, 129.7±3.64mmol/l and 122.7±2.57mmol/l respectively. The values of potassium for HIE stage I, II and III were 4.86±0.67 mmol/l, 5.46±0.78 mmol/l and 6.16±0.83 mmol/l respectively. Similarly, the mean values of calcium for HIE stage I, II and III were 8.06±1.25mg/dl, 7.13±.77 mg/dl, 6.03±0.79 mg/dl respectively. The values of sodium, potassium and calcium among different severity of asphyxia were significantly different (p-value 0.001, 0.02 and 0.01 respectively). The more severity of HIE stages determine the more severe hyponatrimia, hyperkalemia and hypocalcemia.*

Conclusion: *Electrolyte changes occur frequently in asphyxiated neonates. The degree of hyponatremia, hyperkalemia and hypocalcemia was directly proportional to the severity of perinatal asphyxia.*

Key Words: *Perinatal asphyxia, hyponatremia, hyperkalemia, hypocalcaemia*

---

1. Assistant Professor, Department of Pediatrics, Dr. M R Khan Shishu Hospital and Institute of Child Health, Dhaka.
2. Registrar, Department of Pediatrics, Anwer Khan Modern Medical College & Hospital.
3. Associate Professor, Department of Pediatrics, Dr. M R Khan Shishu Hospital and Institute of Child Health, Dhaka.
4. Assistant Registrar, Department of Pediatrics, Dr. M R Khan Shishu Hospital and Institute of Child Health, Dhaka.

Correspondence to: Sheikh Farjana Sonia, Assistant Professor, Department of Paediatrics, Dr. M R Khan Shishu Hospital and Institute of Child Health, Mirpur-2, Dhaka. Cell: 01752038848, Email: soniafarzana7@gmail.com
Received: 15 March 2018; **Accepted**: 16 April 2018

Introduction

Perinatal asphyxia is an important cause of metabolic derangements and acute neurologic injury in newborns. Despite of the important advances in perinatal care in the past decades, asphyxia remains a severe condition leading to significant mortality and morbidity.[1] The term "asphyxia" is derived from the Greek word and means "stopping of the pulse". Perinatal asphyxia is a condition where there is delay in establishing spontaneous respiration upon delivery of a newborn and characterized by an impairment of exchange of the respiratory gases (oxygen and carbon dioxide) resulting in hypoxemia and hypercapnia, accompanied by metabolic acidosis.[2]

Worldwide incidence of perinatal asphyxia is 2-10/1000 live full term births.[3] It has been shown to be the third most common cause of neonatal death (23%) after preterm birth (28%) and severe infections (26%).[4] According to the World Health Organization (WHO), around four million newborns develop perinatal asphyxia annually.[5] Metabolic derangements have been seen in asphyxiated newborns along with multisystem involvement. In neonate, specific symptoms of electrolyte abnormalities often merge with the features of underlying HIE and inappropriate use of fluid and electrolytes in such situation perpetuates morbidity and mortality.[6] The electrolyte imbalance manifests in the form of hyponatremia, hyperkalemia and hypocalcaemia which have significant linear correlation with the severity of perinatal asphyxia.[6-7]

Normally hypernatremia is expected in the early neonatal period as there is contraction of extracellular fluid due to excretion of water through kidney and high insensible water loss whereas in neonates with perinatal asphyxia there might be hyponatremia as there is increased secretion of anti-diuretic hormone (ADH) in neonates with HIE which leads to increased water retention and hence dilutional hyponatremia.[8] The other reason for hyponatremia is that the capacity of sodium reabsorption is limited and if the load of sodium reaching the collecting tubules (CT) increases significantly, reabsorption doesnot occur proportionately and the sodium load is excreted in the urine.[9] Other contributing factors to hyponatermia are partial resistance to aldosterone.[10]

The rise of serum potassium can be explained from the fact that perinatal asphyxia is associated with acidosis, and in metabolic acidosis, more than one half of the excess hydrogen ions are buffered in the cells. In this setting, electro neutrality is maintained in part by the movement of intracellular potassium into the extracellular fluid. It can also be due to acute renal failure secondary to birth asphyxia which leads to decreased excretion of potassium and hence hyperkalemia.

After birth of a newborn, with the abrupt termination of calcium transport across the placenta, plasma calcium falls, reaching a nadir at age 24-48 h.[11] Serum parathyroid hormone (PTH) increases postnatally in response to this fall in plasma calcium concentration. This increase in PTH mobilizes calcium from bone, and plasma calcium concentration rises. Clinically significant hypocalcaemia occurs in asphyxiated newborns.[12] The etiology behind this is a sluggish response in PTH secretion to the postnatal fall in plasma calcium concentration.

Sodium, potassium and calcium are the major electrolytes inhuman body and any deviation from their normal levels in blood might cause convulsion, shock and other types of metabolic abnormalities. While treating seizures, correction of the electrolyte disturbance is more effective than using anticonvulsants.[13] Hyperkalemia is associated with cardiac dysfunction and death. Calcium is an important second messenger in our body and also helps carrying out muscle function and acts as cofactor for several enzymatic activities. Hypocalcaemia is associated with jitteriness, cardiac dysfunction and seizure. Body should maintain optimum level of these electrolytes in blood. Further the degree of electrolyte imbalance may vary according to the severity of perinatal asphyxia.

During the last 3 decades, under-five mortality and infant mortality rate (IMR) have decreased significantly in Bangladesh, but neonatal mortality remains very high. Out of 3.8 million babies born every year in Bangladesh, 150000 babies die in the first 28 days of life, which is equivalent to one newborn in every 3.4 minutes.[14] There is an unacceptably high neonatal mortality in Bangladesh (20.1/1000 live birth). Early recognition and treatment of electrolyte changes are essential for optimal management and satisfactory long-term outcome. Considering these facts, the study was conducted with the aim to determine the electrolyte (sodium, potassium, calcium) changes in asphyxiated newborns of different severity in the early neonatal period and to find out the correlation of levels of sodium, potassium and calcium with the severity of perinatal asphyxia.

Materials and Methods

This cross sectional study was conducted in special care neonatal ward of Dr. M. R. Khan Shishu Hospital & Institute of Child Health, Mirpur-2, Dhaka over a period of six months from September 2017 to February 2018. A total of 120 term neonates with perinatal asphyxia presented within 3 days of life of both sexes were included in the study. Any preterm neonate, IUGR, neonate with congenital malformations, congenital or perinatal infection, clinically suspected for metabolic diseases and patients whose parents refuse to give written consent were excluded from the study. After proper management of airway, breathing and circulation according to standard guidelines, baseline demographic profile and clinical presentations were taken by a structured questionnaire and 3 ml of venous blood were collected for measurement of serum electrolytes and serum calcium level under aseptic precaution. The severity of asphyxia was staged by Sarnat and Sarnat staging. Serum sodium estimation was done by ion selective electrode method. The Serum calcium levels were measured by 'end point calorimetric method' using O cresolphethelin-complexone or OCPC.

All data were recorded systematically in preformed data collection form and quantitative data was expressed as mean and standard deviation. Statistical analyses of the results were obtained by using window based computer software devised with Statistical Packages for Social Sciences (SPSS-16). Statistical tests for significance of difference were done using chi-square (x^2) test and comparison of mean values of different electrolytes with the different severity of perinatal asphyxia was performed by ANOVA test for parametric data. A 'p' value <0.05 was considered as significant.

Results

In this study, among the 120 asphyxiated neonates, 64(53.3%) patients were male newborn and 56(46.7%) patients were female. Male and female ratio was 1.14:1 and mean weight being 2726(±214.58) gram. The mean gestational age in weeks and age in days of the neonates were 38.54(±1.073) weeks and 3.08 (±1.86) days respectively. It was found that 68 newborns (56.7%) were delivered at hospital or Nagar Shasthya Kendra and 52 patients (43.3%) were delivered at home. Most of newborn, 62(51.7%) were delivered by vaginal delivery, 54(45%) had LSCS and 4(3.3%) had assisted delivery by forceps or ventose (Table I).

Table I
General characteristics of neonates with perinatal asphyxia

Patient data	Characteristic or parameter
Gestational Age (Weeks)	38.54(±1.073)
Gender (M/F)	64/56
Age (Days)	3.08 (±1.86)
Birth weight (Gram)	2726(±214.58)
Place of delivery (Home/ Hospital)	52/68
Mode of delivery (NVD/ LUCS/ Assisted delivery)	62/54/4

According to Sarnat and Sarnat staging of perinatal asphyxia, 44(36.7%) neonates fell under HIE stage I, 60(50%) neonates were under HIE stage II and 16 (13.3%) were in HIE stage III. In stage I, 11.4% neonate developed hyponatrimia, 4.5% neonate developed hyperkalemia and 20.5% neonate developed hypocalcemia. In stage II, 41.7% neonate developed hyponatrimia, 33.3% neonate developed hyperkalemia and 35% neonate developed hypocalcemia. In stage III, 75% neonate developed hyponatrimia, 87.5% neonate developed hyperkalemia and 62.5% neonate developed hypocalcemia (Table II).

Table II
Electrolyte status according to different stages of HIE

HIE stage	No	Hyponatremian n (%)	Hyperkalemian n (%)	Hypocalcaemian n (%)
Stage I	44	5 (11.4)	2 (4.5)	9(20.5)
Stage II	60	25 (41.7)	20 (33.3)	21(35)
Stage III	16	12 (75)	14 (87.5)	10(62.5)
Total	120	42 (35)	36 (30)	40(33.3)

Table III
Distribution of patient according to electrolyte abnormalities

Total Patient	Normal electrolyte		Abnormal electrolyte		p value*
	Number of Patient	Percentage	Number of Patient	Percentage	
120	78 (Normal sodium)	65	42 (Hyponatrimia)	35	<0.01
	84 (Normal potassium)	70	36 (Hyperkalemia)	30	0.04
	80 (Normal calcium)	66.7	40 (Hypercalcemia)	33.3	0.02

*χ^2 test was done to measure the level of significance; p value <0.05 was considered as significant

The study showed out of 120 neonates with perinatal asphyxia 35% of subjects (42 patients) had hyponatremia irrespective of the severity of birth asphyxia which was statistically significant (p value <0.01). Around 30% of subjects (36 patients) presented with hyperkalaemia and 33.3% of subjects (40 patients) presented with hypocalcaemia (p value 0.04 and 0.02 respectively) [Table III].

Table IV
Difference of electrolyte values at different stages of HIE

Electrolytes	HIE stage I mean±SD	HIE stage II mean±SD	HIE stage III mean±SD	p value*
Sodium (mmol/l)	135.62±5.51	129.7±3.64	122.7±2.57	0.001
Potassium (mmol/l)	4.86±0.67	5.46±0.78	6.16±0.83	0.02
Calcium (mg/dl)	8.06±1.25	7.13±0.77	6.03±0.79	0.01

* ANNOVA test was done to measure the level of significance; p value <0.05 was considered as significant

The mean serum sodium level in HIE stage I, stage II and stage III were 135.62±5.51 mmol/l, 129.7±3.64 mmol/l and 122.7±2.57 mmol/l respectively. The mean serum potassium level in HIE stage I, stage II and stage III were 4.86±0.67 mmol/l, 5.46±0.78 mmol/l and 6.16±0.83 mmol/l respectively. The mean serum calcium level in HIE stage I, stage II and stage III were 8.06±1.25 mg/l, 7.13±0.77 mmol/l and 6.03±0.79 mmol/l respectively. On comparing the means of sodium, potassium and calcium between different stages of HIE using ANNOVA, there was significant difference between them with p-value 0.001, 0.02 and 0.01 respectively (Table IV).

Discussion

Sodium, potassium and calcium are the major electrolytes in human body and any deviation from their normal levels in blood might cause convulsions and other metabolic abnormalities. Body should maintain optimum level of these electrolytes in blood.[15] Abnormalities in these electrolyte levels may be a risk factor for the brain injury for an already asphyxiated neonate.[16] Knowledge of these abnormalities among asphyxiated newborns is very valuable to the pediatricians as it is an important variable affecting perinatal mortality.[17] Immediate aggressive treatments of these abnormalities could modify the entire outcome of the babies.[18] This study was an attempt to determine the electrolytes changes among patients with perinatal asphyxia and to find out the severity of these abnormalities with the different stages of HIE.

It was observed that babies with asphyxia had higher incidence of hyponatremia. In this study out of 120 asphyxiated babies 42 (35%) were hyponatremic and hyponatremia was the predominant electrolyte abnormality. This finding is consistent with finding of Singhi et al and Prasad et al where hyponatraemia was found in 30% and 29.8% respectively.[19, 20] This observation was also made by Basu et al. where significant hyponatremia was noted (p <0.001).[18] In

another study by Gupta et al observed that asphyxiated babies had lower mean serum sodium levels as compared to the control group (p <0.001).[21] Presence of SIADH in perinatal asphyxia explain high incidence of hyponatremia in these neonates. But these findings were in contrast with findings of Hossain et al where hyponatremia was reported in 26.7% neonates and they also found hypernatremia in 23.8% of asphyxiated neonates which was not found in the present study.[22]

In this study, about 75% of severely asphyxiated, 41.7% of moderately asphyxiated and 11.4% of mildly asphyxiated babies were hyponatremic.This finding describe that fall in serum sodium concentration may be associated with severity of asphyxia. The positive association between serum sodium and staging of HIE was found to be statistically highly significant (P<0.001). This finding is similar to a study done by Basu et al. They interpret that when the severity of HIE stages increasing, the sodium level tends to fall (P <0.01).[18]

In this study, around 30% of subjects (36 patients) presented with hyperkalaemia. Two other study done by Gupta et al and Basu et al higher serum potassium levels were also observed in asphyxiated neonates as compared to those with controls (p <0.001).[18, 21] Lackmann et al, measured potassium levels in 98 asphyxiated new-borns and none of them showed significant hyperkalaemia in the initial 144 hours of life which is contrary to our study.[23] Two other study done by Hossian et al and Marudhkar et al where they found 8.6% and 14.8% cases of hypokalemia in asphyxiated neonates.[22, 24] But in our study we did not find any case of hypokalemia. Singhi et al and Roa et al. found hyperkalemia in 5.4% and 14.4% respectively of ICU admissions, which included asphyxiated as well as other sick neonates which is similar to our study.[25, 26]

In this study, mean serum potassium levels were 4.86±0.67mmol/l in stage I, 5.46±0.78 mmol/l in stage II and 6.16±0.83mmol/l in stage III i.e. the more the degree of HIE the more the level of serum potassium which was statistically significant (p value 0.02). This finding is similar to the study done by Basu et al. They found hyperkalaemia is directly proportionate to the severity of HIE stages with significant p value (P value <0.01).[18]

In this study out of 120 asphyxiated babies, 40 (33.3%) neonates developed hypocalcemia. Similarly in a case control study by Jajoo et al and Rai et al and Schedewie et al showed that asphyxiated newborns had lower serum calcium level compared to their controls.[27-29] Our study determine that there was significant difference of mean serum calcuim level among the three stages of HIE (p value 0.01). The degree of hyocalcemia is directly proportionate to the severity of HIE stages. This finding is similar with the study done by Basu et al where similar to sodium, calcium levels also presented with negative correlation with the HIE stages.[18]

Azmeri et al showed in their study that there was no significant differences of biochemical changes between HIE stage II & HIE stage III.[30] But in the present study it was found that there was significant difference of biochemical parameter among HIE stage I, stage II and stage III.

Our study had some limitations. Our classification of HIE was according to Sarnat and Sarnat stage which is simple but it doesn't take various parameters into consideration like EEG and USG and this study is a single centered study.

Conclusion

The electrolyte changes manifest in the form of hyponatremia, hyperkalemia and hypocalcaemia which are most common associated abnormalities in neonates with perinatal asphyxia. More severe hyponatremia, hypocalcemia and hyperkalemia should be suspected if there is severe perinatal asphyxia and vice versa. Early recognition of this electrolyte changes may reduce morbidity and mortality of neonate with perinatal asphyxia.

References

1. Roberto Antonucci, Annalisa Porcella, Maria Dolores Pilloni et al. Perinatal asphyxia in the term newborn. *Journal of Pediatric and Neonatal Individualized Medicine* 2014; **3**(2): 1-14.
2. Bax MC, Flodmark O, Tydeman C. Definition and classification of cerebral palsy. From syndrome toward disease. *Dev Med Child Neurol Suppl*. 2007;**109**:39-41.
3. Brain damage from perinatal asphyxia: correlation of MR findings with gestational age- Barkovich and Truwit 11 (6): 1087- www.ajnr.org. *Retrieved* 2008; 03-27.
4. Lawn JE, Cousens S, Zupan J and for the Lancet Survival Steering team. 4 million neonatal deaths: when? where? why? *Lancet* 2005; **365**: 891-900.
5. Lawn JE, Wilczynska-ktende K, Cousens SN. Estimating the causes of four million deaths in the

year 2000. *Int J Epidemiol* 2006:**35**(3);718-19.

6. Aggarwal R, Deorari AK, Paul VK. Fluid and electrolyte management in term preterm neonates. *Indian J Pediatr* 2001; **68**: 1139-42.
7. Wei Z, Xia Y, Qing PF, Ting YL.Correlation of brain natriuretic peptide with hyponatremia in newborn infants with hypoxic ischemic encephalopathy.*World J Pediatr* 2007; **3**: 121-24.
8. Bauer K, Versmold H. Postnatal weight loss in preterm neonates <1500g is due to isotonic dehydration of the extracellular volume. *Acta Paediatr* 1989; **78**(s360): 37-42.
9. Wu PY, Hodgman JE. Insensible water loss in preterm infants: changes with postnatal development and non-ionizing radiant energy. *Pediatrics* 1974; **54**(6): 704-12.
10. Shaffer SG, Meade VM. Sodium balance and extracellular volume regulation in very low birth weight infants. *J Pediatr* 1989; **115**(2): 285-90.
11. Tsang RC, Chen IW, Friedman MA, Chen I. Neonatal parathyroid function: Role of gestational age and postnatal age. *J Pediatr* 1973; **83**(5): 728-38.
12. Tsang RC, Kleinman LI, Sutherland JM, Light IJ. Hypocalcemia in infants of diabetic mothers: Studies in calcium, phosphorus, and magnesiummetabolism and parathormone responsiveness. *J Pediatr* 1972; **80**(3): 384-95.
13. Melkie M, Yigeremu M, Nigussie P, Teka T, Kinde S. Establishing reference intervals for electrolytes in newborns and infants using direct ISE analyzer. *BMC Res Notes* 2013; **6**(1): 199.
14. Newborn health in Bangladesh, A situation analysis, Save the children. 2001.
15. Chapman IA, Stoll BJ. Nervous System Disorders. In: Behrman RE, Kliegman RM, Jenson HB, Stanton BF, editors. Nelson Text Book of Pediatrics.18th ed. Philadelphia: WB Saunders Elsevier 2007:713-22.
16. Seyal T, Hanif A. Factors related to adverse outcome in asphyxiated babies. *Ann King Edward Med Uni* 2009; **15**: 80-84.
17. Chawla D, Agarwal R, Deorari AK, Paul VK. Fluid and electrolyte management in term and preterm neonates. *Indian J Pediatr* 2008; **75**(3): 255-59.
18. Basu P, Som S, Das H, Choudhuri N. Electrolyte status in birth asphyxia. *Indian J Pediatr* 2010; **77**: 259-62.
19. Singhi S. Hyponatremia in hospitalized critically iII children: Current concepts. *Indian J Pediatrics* 2004; **71**: 803-07.
20. Prasad SVSS, Singhi S, Chugh KS. Hyponatremia in sick children seeking pediatric emergency care. *Indian Pediatrics* 1994; **31**: 287-94.
21. Gupta BD, Sharma P, Bagla J, Parakh M, Soni JP. Renal failure in asphyxiated neonates. *Indian Pediatrics* 2005; **42**: 928-34.
22. Hossain MM, Shirin M, Mamun AA. Electrolyte Abnormalities in Neonates Admitted in Intensive Care Unit. *Bangladesh J Child Health* 2004; **28**(1): 13-17.
23. Lackmann GM, Mader R, Tollner U. Serum potassium level in healthy neonates and infants with asphyxia in the first 144 hours of life. *Klinische Padiatrie* 1991; **203**(5): 399-402.
24. Marudkar A, Singhi S. Hypokalemia in a pediatric intensive care unit. *Indian Pediatrics* 1996; **33**: 9-14.
25. Singhi S, Gulati S, Prasad SV. Frequency and significance of potassium disturbances in sick children. *Indian Pediatr* 1994; **31**(4): 460-63.
26. Rao SDS, Thomas B. Electrolyte abnormalities in children admitted to pediatric intensive care unit. *Indian Pediatrics* 2000; **37**: 1348-53.
27. Jajoo D, Kumar A, Shankar R, Bhargava V. Effect of birth asphyxia on serum calcium levels in neonates. *Indian J Pediatr* 1995; **62**: 455-59.
28. Rai S, Bhatiyani KK, Kaur S. Effect of birth asphyxia on serum calcium and glucose level: A prospective study. *Int J Sci Stud* 2015; **3**(7): 3-6.
29. Schedewie HK, Odell WD, Fisher DA, Krutzik SR, Dodge M, Cousins L, et al. Parathormone and perinatal calcium homeostasis. *Pediatr Res* 1979; **13**(1):
30. Azmeri S, Ruma P, Shima B, Nobo KG, Kazi I, Sheikh FS, Farhana R. Biochemical changes in asphyxiated neonate presented with convulsion. *BAOJ Pediatrics* 2018; **4**: 2:1-5.

DS (Child) H J 2018; 34(1) : 94-97

ORIGINAL ARTICLE

Effect of Topical Estrogen in Pre-pubertal Labial Adhesion

Gajendra Nath Mahato[1], Paritosh Kumar Palit[2], Md. Hasanuzzaman[3], Md. Moniruzzaman,[4] Md. Abdus Sattar,[5] Gouranga Kumar,[6]

Abstract

Background: *Labial adhesion is the partial or complete adherence of the labia minora originating at the posterior fourchette and extending towards the clitoris. The cause of labial adhesion appears to a mild inflammatory condition in a child with a thin layer of labial epithelial cells secondary to a low estrogen level.*

Objective: *The purpose of the study was to evaluate the effectiveness of topical estrogen in the treatment of labial adhesion in pre-pubertal girls.*

Methods: *This prospective study was carried out among 100 pre-pubertal girls with labial adhesion at the Out Patient Department of Dhaka Shishu (Children) Hospital from January 2014 to December 2016. Patients previously treated with topical estrogen for labial adhesion was excluded from the study. The parents of the children were instructed to applya thin layer of estrogen cream to the raphe along the adhesion line twice daily. All children were treated with 1 to 2 courses. All patients were followed at 3rd week, 6th week, 3rd month, 6th month. During each follow up visit, release of adhesion and adverse effect were noted.*

Results: *The mean age of the pre-pubertal girls was 11.47±5.56 months and mean weight was 8.39±2.04 kg. Among them 45.0% patients had complete adhesion while 52.0% had associated symptoms. Labial adhesion was released within 3 weeks in 71.0% patients and within 6 weeks in 90.0% patients. Within 6 months follow up, recurrence of adhesion was found in 8.9% patients and adverse effect developed in 15.0% patients. Among the patients, success rate was 82.0% with topical estrogen treatment.*

Conclusion: *Topical estrogen is effective and safe in the treatment of labial adhesion in pre-pubertal girls.*

Key words: *Labial adhesion, pre-pubertal girl, topical estrogen.*

Introduction

Labial adhesion is the partial or complete adherence of the labia minora[1] originating at the posterior fourchette and extending towards the clitoris.[2] Most children with minor agglutination of the labia are asymptomatic. When symptoms occur, they are often related to interference with voiding, such as dysuria or altered urinary stream, or symptoms related to the accumulation of urine behind the agglutination predisposing to vaginal or urinary tract infections.[3]

1. Registrar, Division of Pediatric Surgery, Dhaka Shishu (Children) Hospital.
2. Associate Professor, Division of Pediatric Surgery, Shaheed Suhrawardy Medical College Hospital, Dhaka.
3. Registrar, Pediatric Surgery, Shaheed Suhrawardy Medical College Hospital, Dhaka.
4. Resident Medical officer, Division of Pediatric Surgery, Dhaka Shishu(Children) Hospital.
5. Assistant Professor, Department of Pediatric Surgery, Pabna Medical College, Pabna.
6. Junior consultant (surgery) Upazila Health Complex, Barishal, Tangail.

Correspondence to: Dr. Gajendra Nath Mahato, Registrar, Division of Pediatric Surgery, Dhaka Shishu (Children) Hospital. Cell: 01711347134, E-mail: dr.gnmahato@yahoo.com

Received: 15 January 2018; **Accepted**: 12 March 2018

The incidence has been reported as approximately 0.6 to 3.0%.[4] The cause of labial agglutination appears to a mild inflammatory condition in a child with a thin layer of labial epithelial cells secondary to a low estrogen level. When vulvitis or another inflammatory condition occurs, the thinner layer of epithelia cells can denudate and apposition of the eroded areas can result in agglutination of the labia.[5] About 80% of labial adhesions resolve spontaneously within 1 year[6] and persistence after the onset of puberty is very rare. Management options range from reassurance for asymptomatic patients to surgical treatment of severe cases and cases resistant to conservative treatment.[7] Many physicians offer young girls estrogen cream to be applied on the labia for several weeks.The success rate of topically applied estrogen cream ranged between 46.7% and 100%.[8] The present study evaluated the effectiveness of estrogen cream in the treatment of labial adhesion in pre-pubertal girls.

Materials and Methods

This prospective study was carried out on patients who came for treatment of labial adhesion at the Out Patient Department of Dhaka Shishu (Children) Hospital from January 2014 to December 2016. Ethical clearance was taken from Ethical Review Committee of Bangladesh Institute of Child Health & Dhaka Shishu (Children) Hospital and informed written consent was taken from all the parents or legal guardians of the patients after adequately explaining them, the purpose of the study. If the labial adhesion was severe with only a small pin-point opening on the labia, it was termed as complete labial adhesion. If the labial adhesion had a small opening on the labia, it was termed as partial labial adhesion. Patients previously treated with estrogen cream for labial adhesion was excluded from the study. Routine and microscopic examination with culture sensitivity test of urine was done for patients. The parents of the children were instructed to apply a thin layer of Estrogen cream to the raphe along the adhesion line twice daily. A single course of treatment was defined as a 3-week course. All children were treated with 1 to 3 courses. All patients were followed at 3rd week, 6th week, 3rd month, 6th month. During each follow up visit, release of adhesion and adverse effects such as pigmentation of genitalia, breast budding were noted. The statistical analysis was conducted using SPSS (statistical package for social science) version 20 statistical software. Associations of continuous data were assessed using student t- test. Associations of categorical data were assessed using Chi-square test. For both tests, $p<0.05$ was considered significant.

Results

The present study found that the mean age of the patients was 11.47±5.56 months and mean weight was 8.39±2.04 Kg. Among them 45.0% patients had complete adhesion while 52.0% had associated symptoms (Table I). The associated symptoms were UTI (32/52), post void dripping (11/52) and poly urea (9/52) (Figure 1). Labial adhesion was released within 3 weeks in 71.0% patients and within 6 weeks in 90.0% patients (Table II). Within 6 months follow up, recurrence of adhesion was found in 8.9% patients and adverse effect developed in 15.0% patients (Table III). Among the patients, success rate was 82.0% with estrogen cream treatment (Figure 2).

Table 1
Patient's characteristics

Characteristics	
Age (in months)	11.47±5.56
Weight (in Kg)	8.39±2.04
Complete adhesion	45 (45.0%)
Presence of symptom	52 (52.0%)

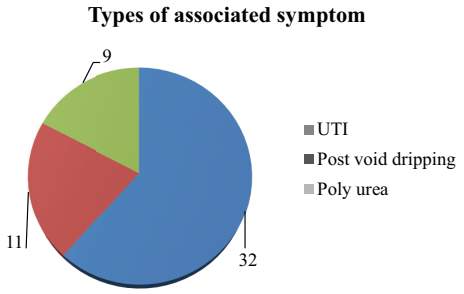

Fig 1 Types of associated symptom (n=52)

Table II
Release of labial adhesion

Release of labial adhesion	No. of patients
Within 3 weeks	71(71.0%)
Within 6 weeks	90(90.0%)

Table III
Recurrence of adhesion and presence of adverse effect within 6 months

Characteristics	No. of patients (%)
Recurrence of adhesion (n=90)	8(8.9)
Adverse effect (n=100)	15(15.0)

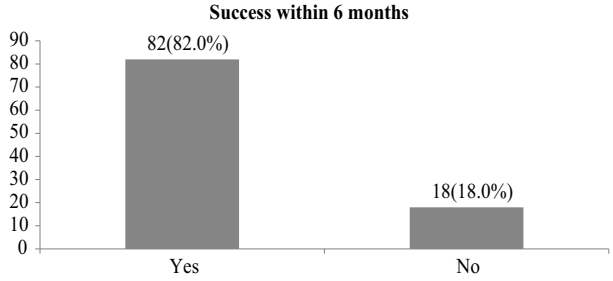

Fig 2 *Success within 6 months*

Discussion

One theory for labial fusion is low pre pubertal estrogen levels. Topical estrogen remains the mainstay of therapy. Estrogen's action in regard to collagen may influence recurrent adhesions and adhesions that form after manual disruption or surgical separation.[1] The present study evaluated the effectiveness of topical estrogen in the treatment of labial adhesion in pre-pubertal girls.

In the present study, the mean age of the patients was 11.47(±5.56) months. Leung et al[5] reported that the peak incidence of labial adhesion was 13-23 months of age which might be a result of the combination of the children's low estrogen level and irritated skin caused by nappy use. He also noticed that the incidence of labial agglutination was lower in children younger than 3 months of age, as they still might be influenced by maternal estrogens.

Although labial adhesion is often asymptomatic, it sometimes results in postvoid dripping, UTIs, vaginitis, hematuria, and urinary frequency. In the present study, 52.0% had associated symptoms other than labial adhesion. Among the patients, 32.0% had UTI. Although UTI occurs in 3-5% of girls in the normal population, the rate in those with labial adhesion can be as high as 20-40%.[9] Melek et al[10] found that there was a marked association between the presence of UTI and the type of adhesions. The percentages of UTIs in girls with complete and partial LA were 84.0% and 28.6%, respectively (P <0.05). The percentages of UTIs in girls with thick and thin LA were 100% and 44.1%, respectively (P <0.05). In the current study 9.0% patients had poly urea and 11.0% had post void dripping. The study of Mayoglou et al[11] found 12.6% patients suffered from post void dripping which matched the present study.

After initiation of treatment, all patients were followed at 3rd week, 6th week, 3rd month and 6th month. Labial adhesion was released within 3 weeks in 71.0% patients. This result was closer with the result of ERTÜRK who found that 65% labial adhesion was released within 3 weeks.[12] Within 6 weeks labial adhesion was released in 90.0% patients. Ten patients had adhesion after 6 weeks. Treatment was stopped for those patients and adhesions were manually released. Within 6 months follow up, recurrence of adhesion was found in 8.9% patients. Other studies found recurrence rate 11.6%, 35% or 41%.[13,11,6] The dissimilarity of result in recurrence rate might be due to the age difference in patients. In the present study, the mean age of patients was 11.48±5.25 months while the patients varied in age from3 months to 10 years in those studies.

Within 6 months follow up, adverse effect developed in 15.0% patients. Short-term side effects of topical estrogens include breast budding, rash or irritation, and vaginal bleeding.[11] In the current study, the side effects of estrogen were found breast budding and hyperpigmentation which developed in 9.0% and 6.0% patients respectively.

The result of the present study indicated that the success rate of the treatment was 82.0%. Different author showed different success rate though in all cases majority of the patients had release of adhesion. A retrospective study was conducted in Turkey in pre pubertal girls where the success rate of the estrogen treatment was 80.0%.[12] Other studies showed success rate 66% and 79% respectively.[14,6] These differences of result might be due to the difference in age of the study population and the type of adhesion.

Conclusion

Topical estrogen is effective and safe in the treatment of labial adhesion in pre-pubertal girls.

References

1. Leung AKC, Robson WLM, Kao CP, Liu EKH, Fong JHS. Treatment of labial adhesion with topical estrogen therapy. *Clin Pediatr* 2005; **44**: 245-47.
2. Valender MH, Mikkelsen DB, Bygum A. Labial agglutination in a prepubertal girl: Effect of topical oestrogen. *Acta Derm Venereol* 2009; **89**: 198-89.
3. Bacon JL. Prepubertal labial adhesions: Evaluation of a referral population. *Am J Obstet Gynecol* 2002; **187**: 327-32
4. Al Jurayyan NAM. Labial agglutination (Adhesion) in pre-pubertal girls. What a primary care physician should kKnow? *Primary Health Care* 2012; **2**: 128.
5. Leung AK, Robson WL, Tay-Uyboco J. The incidence of labial fusion in children. *J Pediatr Child Health* 1993; **29**: 235-36.
6. Schroeder B. Pro-conservative management for asymptomatic labial adhesions in the pubertal child. *J Pediatr Adolesc Gynecol* 2000; **13**: 184-5.
7. Dowlut-McElroy T, Higgins J, Williams K, Strickland J. Treatment of prepubertal labial adhesions: A double blinded, randomized controlled trial comparing topical emollient versus topical estrogen. *J Pediatr Adolesc Gynecol* 2017; **30**(2): 268.
8. Tebruegge M, Misra I, Nerminathan V. Is the topical application of oestrogen cream an effective intervention in girls suffering from labial adhesions? *Arch Dis Child* 2007; **92**(3): 268-71.
9. Sanfilippo JS. Vulvovaginitis. In: Nelson WE, Behrman RE, Kliegman R, editors. Nelson textbook of pediatrics.15th ed. Philadelphia: WB Saunders: 1996.1555.
10. Melek E, K1l1çbay F, Sar1ka N, Bayaz1t A. Labial adhesion and urinary tract problems: The importance of genital examination. *J Pediatr Urol* 2016; 12(2): **111**. e1-111.e5.
11. Mayoglou L, Dulabon L, Martin-Alguacil N, Pfaff D, and Schober J. Success of treatment modalities for labial fusion: A retrospective evaluation of topical and surgical treatments. *J PediatrAdolesc Gynecol* 2009; **22**: 247-50.
12. Erturk N. Comparison of estrogen and betamethasone in the topical treatment of labial adhesions in prepubertal girls. *Turk J Med Sci* 2014; **44**: 1103-07.
13. Muram D. Treatment of prepubertal girls with labial adhesions. *J Pediatr Adolesc Gynecol* 1999; **12**: 67-70.
14. Soyer T. Topical estrogen therapy in labial adhesions in children: Therapeutic or prophylactic? *J Pediatr Adolesc Gynecol* 2007; **20**(4):241-44.

DS (Child) H J 2018; 34(1) : 98-101

ORIGINAL ARTICLE

Pattern of Pediatric & Neonatal Surgical Mortality: Experience at Dhaka Shishu (Children) Hospital

Md Samiul Hasan[1], KM Nur ul Ferdous[1], Umama Huq[2], Md. Abdul Aziz[3], Md. Ayub Ali[1], Ipsita Biswas[4]

Abstract

Background: *Survival after neonatal and pediatric surgery in developed countries are excellent but the situation in developing countries and different.*

Objective: *We wanted to evaluate the outcome of pediatric and neonatal surgical care at our setup and to generate better data for future policy making and measurement of quality improvement.*

Methods: *It was a retrospective analytical study, done in pediatric surgery division, Dhaka Shishu (Children) Hospital, from March 14 to April 17. Data were collected from death register, operation theater register and monthly audit. Patients were divided into 4 groups according to age. Group 1: 0 to 1month, group 2: 1month to 1 year, group 3: 1 year to 5 years, group 4: > 5 years. Information regarding diagnosis, treatment and cause of death were analyzed. Ethical clearance was taken from hospital ethical committee.*

Result: *Total 14411 patients got admitted during this period. Total 9847 (routine 6596 & emergency 3251) operations were performed. Among them 669 (4.64%) patients expired [Male 394 (58.9%) & female 275 (41.1%)]. Among the expired patients 472 (70.6%) were neonates, 157 (23.5%) were between 1 month to 1 year, 31 were between 1 year to 5 year and (9%) patients were >5 years old. Among the neonates 287 (60.8%) were premature. 518 (77.4%) patients had congenital anomalies and 151 (22.6%) patients had acquired conditions. Major preoperative diagnosis was different types of intestinal obstruction (255), abdominal wall defect (84), ARM (58) & intestinal perforation (56). Emergency operation performed in 435 (65%) patients, 116 (17.3%) patients underwent routine operation and the rest 118 (17.6%) patients did not undergo into any surgery. Major cause of death was sepsis (78.7%).*

Conclusion: *Surgery for congenital anomalies at neonatal age as emergency surgery contributed highest mortality. Sepsis was the final cause of death in most of the patient. Improvement of peri operative care is the key to improve the situation.*

Keywords: *Mortality, neonate, pediatric surgery, sepsis*

1. Assistant Professor, Division of Pediatric Surgery, Dhaka Shishu (Children) Hospital, Dhaka.
2. Resident, Division of Pediatric Surgery, Dhaka Shishu (Children) Hospital, Dhaka.
3. Professor, Division of Pediatric Surgery, Dhaka Shishu (Children) Hospital, Dhaka.
4. Associate Professor, Division of Pediatric Surgery, Dhaka Shishu (Children) Hospital, Dhaka.

Correspondence to: Dr Md Samiul Hasan, Assistant Professor, Division of Pediatric Surgery, Dhaka Shishu (Children) Hospital. Cell: 01712886034, E-mail: samiulo45@gmail.com

Received: 5 June 2018; **Accepted**: 21 June 2018

Introduction

Surgical care significantly varies in between countries, institutions and surgeons. Several measures reflect quality of surgical care, among which outcome measurement is most obvious. It is the bottom line of what surgeons do.[1] Measurement of outcome alone can improve the quality of surgical care. Among several outcomes indicators, mortality is an essential indicator of quality of surgical service being provided.[2] Though survival after neonatal and pediatric surgery in developed countries is excellent, situation in developing countries is different. Significantly higher mortality has been reported in developing countries.[3,4] In Bangladesh pediatric surgery has evolved as a subspecialty of surgery over the past decades, though super-specializations within paediatric surgery are yet to come.With limited resource and lots of limitations this subject is thriving throughout the country. To improve further there is no alternative of outcome analysis. In our country, the accurate data containing paediatric surgical patient load, morbidity and mortality is not available. No such study has been performed till this date. The aim of this study was to evaluate the pattern of mortality in paediatric surgical patient in our country and to generate baseline data for future policy making and measurement of quality improvement.

Materials and Methods

It was a retrospective observational study done in paediatric surgery division, Dhaka Shishu (Children) Hospital, from March 2014 to April 2017. Data were collected from death register, operation theater register and monthly audit. All patients expired during this period were included in this study. Patients were divided into 4 groups according to age. Group 1: age 0 to 1month, group 2: 1month to 1 year, group 3: 1 year to 5 years, group 4: more than 5 years.Information regarding diagnosis, treatment and cause of death were analyzed. SPSS version 22 was used for statistical analysis. Ethical clearance was taken from hospital ethical committee.

Results

Total 14411 patients got admitted within this period (Figure 1). Total operation performed during this period is 9847 (routine operation were 6596 and emergency operations were 3251). Total 669 (4.64%) patients expired among which 394(58.9%) were male and 275 (41.1%) were female. 472 (70.6%) were neonates out expired patients, 157(23.5%) were between 1 month to 1 year, (31%) (were between 1 year to 5 year and (9%) patients were >5 years old (Fig 2)

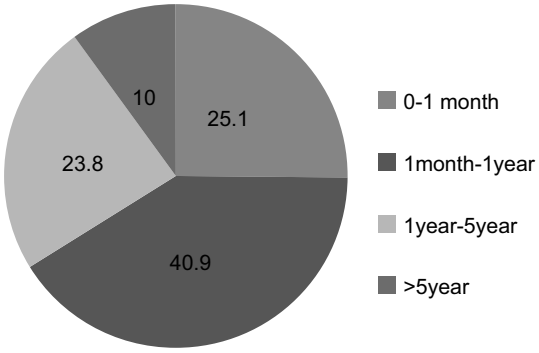

Fig 1 *Age distribution of admitted patients*

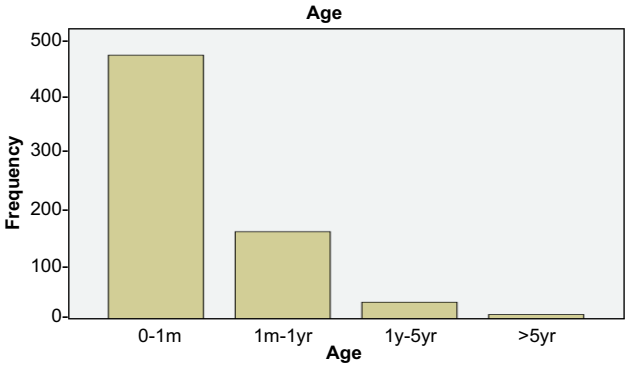

Fig 2 *Age distribution of expired patients*

Among the expired neonates 287 (60.8%) were premature. Emergency operation performed in 435 (65%) patients, 116 (17.3%) patients underwent routine operation and no operation was performed in the rest 118 (17.6%) patients (Figure 3). Majority of expired patients, which is (77.4% /518), admitted with congenital anomalies and 151 (22.6%) patients were with acquired conditions (Table I).

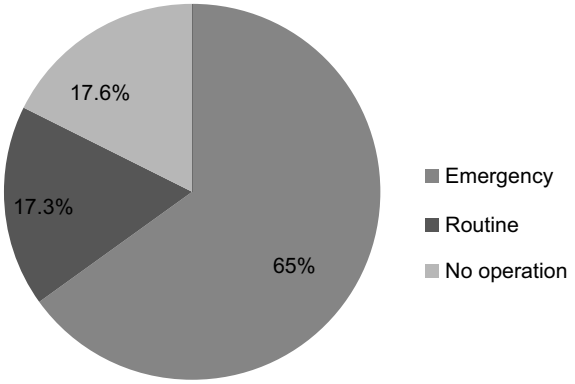

Fig 3 *Type of operation among expired patients*

Table I
Type of surgery among congenital and acquired surgical problems

		Congenital	Acquired	Total
Types of operation	Emergency	354	81	435
	Routine	86	30	116
	Nonoperative	78	40	118
Total		518	151	669

Major preoperative diagnosis was different types of intestinal obstruction (255), followed by abdominal wall defect (84), ARM (58) and intestinal perforation (56) (Figure 4). Major cause of death was sepsis (78.7%). (Figure 5).

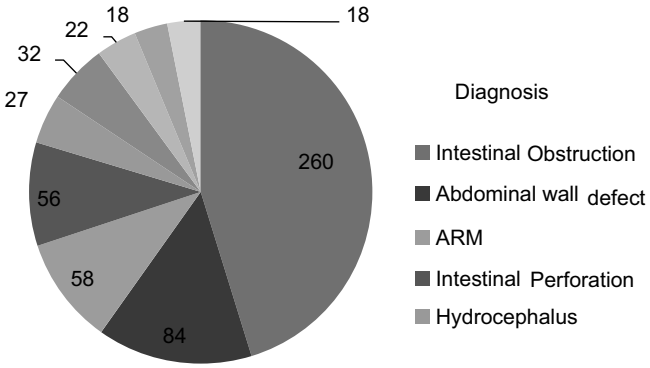

Fig 4 *Distribution of preoperative diagnosis*

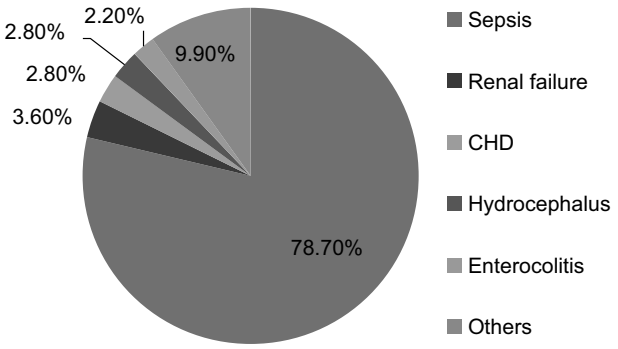

Fig 5 *Causes of death*

Discussion

Dhaka Shishu (Children) Hospital paediatric surgery division is the largest paediatric surgery center of Bangladesh in terms of hospital bed, admitted patients and operations performed. We don't have separate neonatal surgery ward or surgical ICU. Neonatal age group contributed highest mortality followed by infants. Most paediatric surgical mortality studies reported this finding.[3,4] This is may be due to physiological immaturity and vulnerability to sepsis of these young patients. Overall mortality (4.64%) and neonatal surgical mortality (13%) both are much lower in comparison to other studies reported from developing countries but much higher from developed world.[3-6] The improved survival in developed country is not due to improved surgical skill rather due to growth of pediatric surgery andanesthesia, better understanding of neonatal physiology, sophisticated technology that allows better monitoring & intensive care, improved airway management, Total parenteral nutrition (TPN) and effective antibiotics.[5]

Emergency surgery and surgery for congenital anomalies showed significantly higher mortality in developing countries worldwide as more than 90% of congenital anomalies affect low income countries.[3,7-9] Studies form developing countries identified several factors influencing this high mortality rate. Khan et al[10] described unplanned pregnancy and delivery, failure to maintain asepsis, lack of sophisticated equipmentand monitoring system as the determining factors. Ilori et al[11] found the three delay model (delay in recognition of illness, delay in accessing care & delay in provision of care) responsible for higher mortality. Poverty, illiteracy, cultural beliefs, inadequate health care facilities in rural areas and poor transportation and referral system are responsible for these delay.

In our center, specialist paediatric surgeons are available round the clock but delayed presentation makes the situation worse which is reflected by the percentage of patient died before operation (17.6%). In these patients, disease processes were already far advanced when they reached hospital and succumbed during resuscitation. Ekenze et al.[12] documented 'delayed presentation and inadequate facilities' as the

major challenge in neonatal surgery in developing country.

Sepsis was found as the major cause of death in this study. This is reflected in most of the mortality studies.[11,13,14] Prematurity and infection were found among the three most common causes of neonatal mortality in Bangladesh.[15] Inadequate antenatal care, unhygienic delivery, poor transportation, crowded hospitalward and inadequate infection control practices are responsible for high mortality developing countries as well. Mitul et al[16] described sepsis as the final common pathway to death in a critically ill patient who suffers from a postoperative complication or requires long term mechanical ventilation and TPN. Manchanda et al concluded that adequate antenatal care and asepsis during transfer & handling of the babies will improve the prognosis of surgical neonates.[6] Bhatnagar et al[17] paid importance on awareness of health care professionals, improvement of surgical NICU facilities and detailed study on incidence and causation of sepsis in surgical neonates.

Despite poor economical structure, Bangladesh has done exceptionally well in achieving Millennium Development Goal (MDG) in terms of reducing under five mortality[15]. Without paying proper attention to the surgical mortality, it will be very difficult to achieve Sustainable Development Goal (SDG), which aims to end preventable neonatal death.[18]

Conclusion

Emergency surgery for congenital anomalies at neonatal age contributed highest mortality as per this study. Sepsis and prematurity were the commonest cause of death. Awareness in antenatal diagnosis and early treatment, improvement of NICU facilities and control of sepsis can improve the situation and will pave the way to achieve SDG.

References

1. Birkmeyer JD, Dimick JB, Nancy JO. Measuring the Quality of Surgical Care: Structure, Process, orOutcomes? *J Am Coll Surg* 2004; **198(**4): 626-32.
2. Fitzgerald TN, Moss RL. Clinical outcomes evaluation and quality improvement. In: Coran AG, AdzickNS, Krummel TM, Laberge J, Shamberger RC, Caldamone AA.(eds). Pediatric Surgery, 7th edition. Elsevier, Philadelphia, 2012; 227-36.
3. Global Surg Collaborative. Determinants of morbidity and mortality following emergency abdominal surgery in children in low-income and middleincome countries. *BMJ Global Health,* 2016; **1**: e000091. doi:10.1136/bmjgh-2016-00009.
4. Livingston MH, DCruz J, Pemberton J, Ozgediz D, Poenaru D. Mortality of pediatric surgical conditions inlow and middle income countries in Africa. *Journal of Pediatric Surgery* 2015; **5:** 760-64.
5. Rowe MI, Rowe SA. The last fifty years of neonatal surgical management. *Am J Surg* 2000; **180**: 345-52.
6. Manchanda V, Sarin YK, Ramji S. Prognostic factors determining mortality in surgical neonates. *J Neonatal Surg* 2012; **1**: 3.
7. Lawn JE, Blencowe H, Oza S, You D, Lee AC, Waiswa P, et al. Every Newborn: progress, priorities, and potential beyond survival. *Lancet* 2014; **384**: 189–205.
8. Ameh EA, Dogo PM, Nmadu PT. Emergency neonatal surgery in a developing country. *Pediatr Surg Int.* 2001; **17**: 448-51.
9. Ugwu RO, Okoro PE. Pattern, outcome and challenges of neonatal surgical cases in a tertiary teaching hospital. *Afr J Paediatr Surg* 2013; **10**(3): 226-30.
10. Khan N. On reducing mortality and morbidity of neonatal surgical care. *J Neonat Surg* 2013; **2**: 1.
11. Ilori AM, Ituen CSE. Factors associated with mortality in neonatal surgical emergencies in a developing tertiary hospital in Nigeria. *Open Journal of Pediatrics* 2013; **3**: 231-35.
12. Ekenze SO, Ajuzieogu OV, Nwomeh BC. Challenges of management and outcome of neonatal surgery in Africa: a systematic review. *Pediatr Surg Int* 2016; **32**(3):291-99.
13. Osifo O, Oriaifo IA. Factors affecting the management and outcome of neonatal surgery in Benin City, Nigeria. *Eur J Pediatr Surg* 2008; **18**: 107-10.
14. Ameh EA, Dogo PM, Nmadu PT. Emergency neonatal surgery in a developing country. *Pediatr Surg Int* 2001; **17**: 448-51.
15. Halim A, Dewez JE, Biswas A, Rahman F, White S, van den Broek N. When, Where, and Why Are Babies Dying? Neonatal Death Surveillance and Review in Bangladesh. *PLoS ONE 2016;* **11**(8): e0159388.
16. Mitul AR. Surgical neonatal sepsis in developing countries. *J Neonat Surg* 2015; **4**:41.
17. Bhatnagar SN, Sarin YK. Current trends in neonatal surgery in India. *J Neonatal Surg* 2012; **1**:18.
18. United Nations Development Programme (UNDP). SUSTAINABLE DEVELOPMENT GOALS/ Goal 3: Good health and well being. Available from: URL: http://www.bd.undp.org/content/bangladesh/en/home/sustainable-development-goals/goal-3-good-health-and-well-being.html.

DS (Child) H J 2018; 34(1) : 102-107

ORIGINAL ARTICLE

Pattern of AFP Cases Admitted in a Tertiary Care Paediatric Hospital

Md. Rafiqul Islam[1], Mohammed Rizwanul Ahsan[2], Sabrina Makbul[3], Khaleda Islam[4], Rowshan Kabir[5], ABM Mahfuj Hassan Al Mamun[6]

Abstract

Introduction: *Bangladesh began a comprehensive plan for Acute Flaccid Paralysis (AFP) surveillance from 1998. Now a days identification of pattern of AFP cases is the biggest challenge to take action.*

Objective: *Objective of this study was to detect the major causes of Acute Flaccid Paralysis (AFP) in post polio era and to take necessary action.*

Methods: *This retrospective descriptive study was carried out at Dhaka Shishu (Children) Hospital during the period from January 2017 to July 2017. Data of Acute Flaccid Paralysis were collected including age, gender, laboratory findings and diagnosis.*

Results: *Among 32 non polio AFP cases majority were male 22 (68.75%) and female 10(31.25%). Male and female ratio was 2.2:1. Majority respondents were from 5 year to 10 year aged 16 (50%), < 5 year aged 12 (37.5%) and < 15 year aged 4 (12.5%). Most of respondents had Guillain Barre Syndrome (GBS) 27 (84.4%), Transverse Myelitis 3 (9.4%), Hemiplegia 1(3.1%) and Myositis 1(3.1%).*

Conclusion: *Every case of AFP is a medical emergency. AFP surveillance is the key of early diagnosis and management of AFP case is important. In Bangladesh Guillain Barre syndrome (GBS) is most common cause of AFP. School boys are affected mostly.*

Key words: *Pattern, AFP.*

Introduction

Acute Flaccid Paralysis (AFP) frequently includes respiratory and bulbar weakness. The weakness usually progresses to maximum within days to weeks. Acute Flaccid Paralysis (AFP) is complex clinical syndrome defined as "the acute onset of focal weakness or paralysis characterized as flaccid (reduced muscle tone)".[1] The term "flaccid" indicates the absence of spasticity or other signs of disordered central nervous system motor tracts such as hyperreflexia, clonus, or extensor plantar responses.[2] AFP is related to infectious, traumatic, metal toxicity, or post infectious autoimmune conditions.[3,4] Poliomyelitis was the leading cause of AFP in the prevaccine era.[5] Children 0-4 years of age and unvaccinated adolescents affected mostly.[6] In fact, 99% of Polio Virus infections are mild, show few symptoms, and often go unrecognized.[7] So most Polio Virus infected people are unaware that they carry the virus, thus Polio Virus spreads widely before cases arise. For this reason, in 1996, World Health Organization (WHO) extended polio surveillance to all AFP cases,

1. Assistant Professor, High Deficiency and Isolation (HD & I), Dhaka Shishu (Children) Hospital.
2. Resident Physician & Assistant Professor, Department of Adolescent Medicine, Dhaka Shishu (Children) Hospital.
3. Senior Nutritionist, Dhaka Shishu (Children) Hospital.
4. Professor, Institute of Nutrition and food science, Dhaka University.
5. Institute of Nutrition and Food Science, Dhaka University.
6. Resident Medical Officer, Dhaka Shishu (Children) Hospital.

Correspondence to: Md. Rafiqul Islam, Assistant Professor, High Deficiency and Isolation (HD & I), Dhaka Shishu (Children) Hospital. Cell: 01711850820, Email: rafique_i80@yahoo.com

Received: 13 March 2018; **Accepted**: 26 April 2018

including Guillain-Barré Syndrome (GBS), traumatic neuritis, myelitis/encephalitis, other Enterovirus (EV) infections, and several paralytic syndromes.[7] AFP surveillance continues to be a critical component of the WHO global polio eradication campaign.[8] WHO estimates a background annual incidence of at least 1 rate of case of AFP per 100,000 populations less than 15 years old, in the absence of wild poliovirus transmission.[9,10] Non polio enteroviruses can cause a polio like paralytic disease. Disease progression is slower in paralytic rabies.[11] Herpes group of viruses can lead to AFP by triggering GBS or transverse myelitis, causing polyradiculoneuropathies in immunocompromized hosts.[12] Transverse Myelitis, Traumatic paresis[13] and Hypokalemic Paralysis can also lead to AFP. Worldwide, GBS is considered the most common cause of Acute Flaccid Paralysis in the post-poliomyelitis eradication era.[14-23] It affects people in various geographical locations and virtually all age groups.[24] On the basis of clinical and other information routinely collected through the surveillance system, we defined a GBS case as presence of an Acute Flaccid (hypotonic) Paralysis and symmetrical weakness[18] in the absence of injury or birth trauma. Bangladesh has seven administrative divisions with sixty four districts. Total population of the country as of 2016 is 162,903,941. Approximately 34.3% population is under 15 years of age.[25] Immunization in Bangladesh is performed through fixed and outreach sites with the help of health assistants (HA) and at times family welfare assistants (FWAs) to assist them.[26] WHO has set certain performance indicators for AFP surveillance.[27,28] Bangladesh has been certified as a polio free country with ten other WHO SEAR regions in 2014. The country till 2014 has observed 21 NIDs for polio among which the last one held in January 2014 reached a confirmed coverage of 100%.[29] Bangladesh has successfully made the switch from tOPV to IPV and has withdrawn tOPV from its immunization program following the "Polio Eradication and Endgame Strategic Plan".[30] Patients with AFP classically have motor abnormalities, such as muscle weakness and altered deep tendon reûexes, but tend not to have sensory dysfunction.[31-32] CSF study assist in diagnosis of AFP cases. Magnetic resonance imaging (MRI) has classic findings for infectious anterior myelitis and can assist in establishing the diagnosis.[33] Most causes of AFP include Guillain Barre Syndrome(GBS), transverse myelitis, traumatic neuritis, spinal cord compromise (low back trauma, abscesses or tumors), meningitis, encephalitis, CVA, myopathies, neuropathies and hypokalemic periodic paralysis.[34] This study describes the differential diagnosis of potential causes of AFP, including distribution by age and gender.

Materials and Methods

Retrospectively, we extracted all notified AFP case data from Dhaka Shishu (Children) Hospital Center from January 2017 to July 2017. Causes of AFP were sorted out with their frequencies. All the patients with lower limb weakness were admitted for work-up. Although the initial diagnosis was based upon clinical presentation, few laboratory investigations were carried out on the basis of the clinical picture. Immediately after case notification Two stool specimens collected ≥24 hours apart, both within 14 days of paralysis onset and 100% sample sent through ice or frozen packs to WHO accredited National Polio & Measles Laboratory (NPML) for investigation within 72 hours, arriving in good condition, as per guidelines. Bangladesh tightly follow AFP surveillance set up by WHO guidelines. All the stool samples were sent to virology laboratory for isolation of polio virus. CSF examination was done in most of the cases. A raised CSF protein with normal cell count (albumino-cytological dissociation) suggests Guillain Barre Syndrome. Serum electrolytes, Cerebrospinal fluid examination, Magnetic resonance imaging (MRI) of brain or spine and electrophysiological studies were carried out in certain cases when it was required or diagnosis was doubtful under the guidance of pediatric consultant. The final diagnosis was based on the available clinical data, vaccination history and epidemiology. The surveillance database about AFP cases < 15 years reported during January 2017-July 2017 was used. Data were statistically analyzed with the use of the Statistical Package for Social Science program (SPSS) version 15.0 for windows, Chicago, IL). The descriptive statistical analysis of the quantitative variables was carried out by manual calculation.

Results

Out of 32 non polio AFP cases majority were male 22 (68.75%). Male and female ratio was 2.2:1(Fig-1).

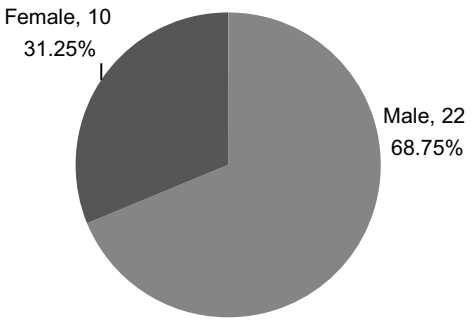

Fig 1 Pie diagram shows demonstration of respondents by gender.(N=32)

Majority, respondents were from 5 year to 10 year < 5 year 12 (37.5%) and <15 year 4 (12.5%)(Fig-2).

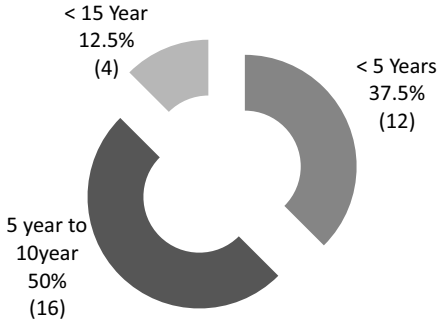

Fig 2 Doughnut diagram showing respondents according to age. (N=32)

After final Diagnosis, most of respondents had GBS 27 (84.4%), Transverse Myelitis 3 (9.4%), Hemiplegia 1(3.1%) and Myositis 1(3.1%) (Fig-3).

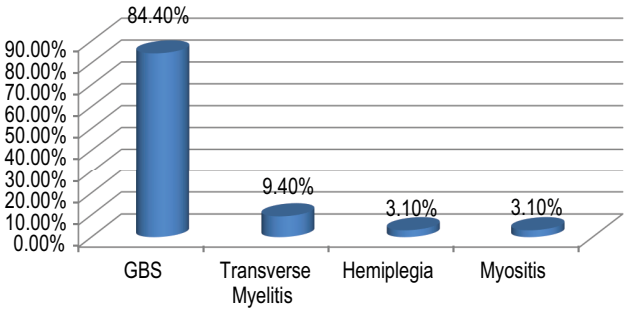

Fig 3 Bar diagram shows respondents according to diagnosis (N=32)

Discussion

In post-polio era, Target of non-polio AFP rate was set to 2/100,000 under 15 years age children[35] but from 2001 till 2015 annualised non-polio AFP rate has always been well over 2 in this country. In this study majority were male 22 (68.75%) and female 10 (31.25%) where male and female ratio was 2.2:1. There is male predominance as seen in other studies conducted in South East Asian region.[37,38] Another study done in Bangladesh by Habib et al[39] found higher incidence of AFP among boys (59.22%) than in girls (40.78%). Similar findings observed in other studies as well.[39-40]

This study has identified that 37.5% of reported AFP were from <5 years of age. Which is similar to data available of Marches region of Italy another country of WHO EURO region 37% of total reported AFP cases belong to under 5 years.[39]

Other studies found,[22,39]more than half of the AFP cases were in children ≤5 years of age. In this study 50% of total reported AFP were from 5 years to 10 years of age, which is similar to a study from Amritsar, India.[36] These findings is not similar to most of the studies but this result may be due to as this study is done in a single centre tertiary based hospital.

With the control of polio, Guillain-Barre Syndrome (GBS) still remains the leading cause of AFP in developed as well as developing countries.[37] This syndrome occurs throughout the world with an annual incidence of GBS ranged from 0.4 to 4 (Mean 1.3) cases per 100 000 per year population.[41-48] This study found 84.4%(27) respondents had GBS, which is slightly higher than the findings of other studies.[49-53] This has been highlighted by the previous studies where incidence of GBS leading to AFP has ranged from 47.3-72.2%.[34,43,54] A study carried out by Jasem et al in Iraqi children identified GBS as a common cause accounting to be 52%. In another study, conducted in Sindh by Memon et al[55] the most common cause of non-polio AFP identified was GBS (21%). Saraswathy et al conducted a study in Malaysia, where GBS was 32.2 %.[23] In this study we found Transverse Myelitis 3 (9.4%) cases. Other studies,[56,57] found transverse myelitis (19%) were higher than that in our study. Similary, Transverse myelitis was the second diagnosis in number and Myositis were among the less frequent diagnoses in other studies.[52,53,58,59]

Conclusion

In Post polio era, AFP is a broad clinical entity with an array of diagnostic possibilities. A systematic

approach helps to narrow down the diagnostic possibilities. Study confirms Guillain-Barre syndrome (GBS) as the most common cause of AFP in the <15 years population, in Bangladesh and most of the countries of the world. This study found school aged boys were mostly suffered by AFP. Irrespective of the cause, generalized weakness frequently affects respiratory and bulbar function so such children need to be managed for respiratory & bulbar muscle weakness and should exclude other causes. Accurate and early diagnosis of the cause has important bearing on the management and prognosis. AFP case based surveillance activity is the key to identify the pattern of AFP cases and to take necessary action.

References

1. Alberta Health and Wellness. Public Health Notifiable Diseases Management Guidelines; August 2011.
2. Growdon JH, Fink JS. Paralysis and movement disorder. In: Isselbacher KJ, Braunwald E, Wilson JD, eds. Harrison's principles of Internal Medicine. New York, NY: McGrawHillBook Company, 1994: 115-25.
3. Marx A, Glass JD, Sutter RW. Differential diagnosis of acute flaccidparalysis and its role in poliomyelitis surveillance. *Epidemiol Rev* 2000; **22**: 298-316.
4. Laxmivandana R, Yergolkar P, Gopalkrishna V, Chitambar SD.Characterization of the non-polio enterovirus infections associatedwith acute flaccid paralysis in south-western India. *PLoS One* 2013; **8**: e61650.
5. Sabin AB. Paralytic consequence of poliomyelitis infection in different parts of the World and I different population group. *Am J Public Health* 1951; 4L: 1215-30.
6. Krugman S, Katz S, L, Gershon AA, Wilfert CM. Enteroviral Infections. *Infectious Diseases of Children:* 1992; **9**: 68-85.
7. World Health Organization (WHO). Weekly Epidemiological Record Relave Epidemiological Ebdomadaire - Acute Flaccid Paralysis (AFP) surveillance: the surveillance strategy of poliomyelitis eradication. 1988: 113-20.
8. Ahmad A, Rehman A. One year surveillance data of acute flaccid paralysis at Bahawalpur Victoria Hospital Bahawalpur. *Pak J Med Sci* 2007; **23**: 308-12.
9. Alcala H. The differential diagnosis of poliomyelitisand other acute flaccid paralysis. *Bio Med Infant Mex* 1993; **50**:136-44.
10. McGrogan A, Madle GC, Seaman HE, de Vries CS. The epidemiology of Guillain Barré syndrome worldwide. A systematic literature review. *Neuroepidemiology* 2009; **32**: 150-163.
11. Gadre G, Satishchandra P, Mahadevan A, et al. Rabies viral encephalitis: clinical determinants in diagnosis with special reference to paralytic form. *J Neurol Neurosurg Psychiatry* 2010; **81**: 812–20.
12. Tyler KL. Herpes simplex virus infections of the central nervous system: encephalitis and meningitis, including Mollaret's. *Herpes* 2004; **11**: 57A-64A.
13. WHO. Report of Interim meeting of Technical advisOly group on the EPI and polio eradication in Western Pacific region. Beijing. Report series no RS/ 1994/ GE/40.
14. Oomes PG, Jacobs BC, Hazenberg MP, Banffer JR, van der Meche FG. Anti-GM1 IgG antibodies and Campylobacter bacteria in Guillain-Barre syndrome: evidence of molecular mimicry. *Ann Neurol* 1995; **38**(2): 170-175.
15. Sladky JT. Guillain-Barre syndrome in children. *J Child Neurol* 2004; 19(3): 191–200.
16. Islam Z, Jacobs BC, van Belkum A, Mohammad QD, Islam MB, Herbrink P, et al. Axonal variant of Guillain-Barre syndrome associated with Campylobacter infection in Bangladesh. *Neurology* 2010; **74**(7): 581-587.
17. Lam RM, Tsang TH, Chan KY, Lau YL, Lim WL, Lam TH, et al. National Committee for the Certification of Wild Poliovirus Eradication: Surveillance of acute flaccid paralysis in Hong Kong: 1997 to 2002. *Hong Kong Med J* 2005; **11**(3): 164-173.
18. Landaverde JM, Danovaro-Holliday MC, Trumbo SP, Pacis-Tirso CL, Ruiz-Matus C. Guillain-Barre syndrome in children aged <15 years in Latin America and the Caribbean: Baseline rates in the context of the influenza A (H1N1) pandemic. *J Infect Dis* 2010; **201**(5):746-750.
19. Molinero MR, Varon D, Holden KR, Sladky JT, Molina IB, Cleaves F: Epidemiology of childhood Guillain-Barre syndrome as a cause of acute flaccid paralysis in Honduras: 1989-1999. *J Child Neurol* 2003; **18**(11): 741-747.
20. Morris AM, Elliott EJ, D'Souza RM, Antony J, Kennett M, Longbottom H. Acute flaccid paralysis in Australian children. *J Paediatr Child Health* 2003; **39**(1): 22-26.

21. Oostvogel PM, Spaendonck MA, Hirasing RA, van Loon AM. Surveillance of acute flaccid paralysis in The Netherlands, 1992-94. *Bull World Health Organ* 1998; **76**(1): 55-62.

22. Poorolajal J, Ghasemi S, Farahani LN, Hosseini AS, Bathaei SJ, Zahiri A. Evaluation of acute flaccid paralysis in hamadan, iran from 2002 to 2009. *Epidemiol Health* 2011; **33**: e2011011.

23. Saraswathy TS, Zahrin HN, Apandi MY, Kurup D, Rohani J, Zainah S, et al. Acute flaccid paralysis surveillance: looking beyond the global poliomyelitis eradication initiative. Southeast Asian *J Trop Med Public Health* 2008; **39**(6):1033-39.

24. Chroni E, Papapetropoulos S, Gioldasis G, Ellul J, Diamadopoulos N, Papapetropoulos T. Guillain-Barre syndrome in Greece: seasonalityand other clinico-epidemiological features. *Eur J Neurol* 2004; **11**(6): 383-388.

25. Global Alliance for Vaccines and Immunisation, Bangladesh. 2016, GAVI.

26. Habib KR, Evaluating Immunisation Dropout Rates in Eight Hard to Reach Unions of Moulvibazar District, Bangladesh 2016.

27. World Health Organisation Field Guide: For Supplementary Activities aimed at achieving polio eradication-1996 revision.1997.

28. CDC Updated guidelines for evaluating public health surveillance systems. Recommendations from the Guidelines Working Group. 2001.

29. World Health Organisation, Bangladesh: home-grown solutions helped stop polio and keep this country polio-free. 2014, World Health Organisation: Bangladesh.

30. World Health Organisation, Polio Eradication & Endgame Strategic Plan 2013-2018. 2013, World Health Organisation:France.

31. Chen F, Li JJ, Liu T, Wen GQ, Xiang W. Clinical and neuroimaging features of enterovirus 71 related acute flaccid paralysis in patients with hand-foot-mouth disease. *Asian Pac J Trop Med* 2013; **6**: 68-72.

32. Messacar K, Schreiner TL, Maloney JA. A cluster of acute flaccid paralysis and cranial nerve dysfunction temporally associated with an outbreak of enterovirus D68 in children in Colorado, USA. *Lancet* 2015;385:1662-71.

33. Chen F, Liu T, Li J, Xing Z, Huang S, Wen G. MRI characteristics and follow-up findings in patients with neurological complications of enterovirus 71-related hand, foot, and mouth disease. *Int J ClinExp Med* 2014; **7**: 2696-704.

35. Tsang RS, Valdivieso-Garcia A. Pathogenesis of Guillian syndrome. *Expert Rev Anti Infec Ther*. 2003; **1**: 597-608.

35. CDC, Progress Toward Interruption of Wild Poliovirus Transmission - Worldwide, January 2005-March 2006. 2005: Washington, DC.

36. Narang G S, Pahwa J S. Retrospective Study of Acute Flaccid Paralysis Cases from a Tertiary Care Centre in Amritsar. *Pediatric Oncall [serial online]* 2011 [cited 2011 February 1]; 8. Art # 14.

37. Jasem J, Marof K, Nawar A, Khalaf Y, Aswad S, Hamdani F, Guillain-Barre Syndrome as a cause of acute flaccid paralysis in Iraqi children: a result of 15 years of nation-widestudy. *BMC neurol* 2013; 10; **13**(1): 195.

38. Habib KR. Polio Eradication in Bangladesh: Evaluation of AFP Surveillance Indicators, 2011-2015; *International Journal of Immunology* 2017;**5**(1):11-19.

39. D'Errico MM, Barbadoro P, Bacelli S, Esposto E, Moroni V, Scaccia F. AFP Study Group. Surveillance of acute flaccid paralysis in the Marches region (Italy): 1997-2007. *BMC Infect Dis* 2008; 8: **135**;

40. Odoom JK, et al., Evaluation of AFP surveillance indicators in polio-free Ghana, 2009–2013. *BMC public health* 2014; **14** (1): p. 1.

41. Kuwabara S. Guillain-Barre syndrome: epidemiology, pathophysiology and management. *Drugs* 2004; **64**(6): 597-610.

42. Molinero MR, Varon D, Holden KR, et al. Epidemiology of childhood Guillain-Barré Syndrome as a cause of acute flaccid paralysis in Honduras: 1989-1999. *J Child Neurol* 2003; **18**(11): 741-47.

43. Barzegar M, Alizadeh A, Toopchizadeh V. Association of Campylobacter jejuni infection and GuillainBarre syndrome: a cohort study in the northwest of Iran. *Turk J Pediatr* 2008; **50**(5): 443-48.

44. Hughes RA, Rees JH. Clinical and epidemiologic features of Guillain-Barre syndrome. *J Infect Dis* 1997; **176** Suppl 2: S92-S98.

45. McGrogan A, Madle GC, Seaman HE, et al. The epidemiology of Guillain-Barre syndrome world wide. Asystematic literature review. *Neuroepidemiology* 2009; **2**(2): 150-63.

46. Prevots DR, Sutter RW. Assessment of Guillain-Barré syndrome mortality and morbidity in the United States: implications for acute flaccid paralysis surveillance. *J Infec Dis* 1997; **175** (Suppl 1): S151.

47. Arami M, Yazdchi M, Khandaghi R. Epidemiology and characteristics of Guillain -Barre syndrome in the northwest of Iran. *Ann Saudi Med* 2006; **26**(1): 22-27.

48. Esteghamati A, Gouya MM, Keshtkar AA, et al. Relationship between occurrence of Guillain-Barre syndrome and mass campaign of measles and rubella immunization in Iranian 5-14 years old children. *Vaccine* 2008; 26(39): 5058-61.

49. Davarpanah M, Bakhtiari H, Mehrabani D,et al. A 12 years surveillance of poliomyelitis and acute flaccid paralysis in Fars Province, Southern Iran. I*ran Red Crescent Med J* 2008;**10**(4): 288-93.

50. Whitfield K, Kelly H. Using the two-source capturerecapture method to estimate the incidence of acute flaccid paralysis in Victoria, Australia. B*ull World Health Organ* 2002; **80**(11): 846-51.

51. D'Souza R, Kennett M, Antony J, et al. Acute flaccid paralysis surveillance in Australia progress report 1995-1998. *Commun Dis Intell* 1999;**23**(5):128-31.

52. Varughese P. Acute flaccid paralysis, Canadian Paediatric Surveillance Program - 2003 Results. Canadian Paediatric Society (Public Health Agency of Canada). Available at: http://www.phac-aspc.gc.ca/publicat/cpsp-pcsp03/page4-eng.php. Access date: Jun 10,2012.

53. Hussain IH, Ali S, Sinniah M, et al. Five-year surveillance of acute flaccid paralysis in Malaysia. *J Paediatr Child Health* 2004;**40**(3):127-30.

54. Rehaman A, Idris M, Elahi M, Arif A. Guillian Barre Syndrome, the leading cause of Acute flaccid paralysis in Hazara division. *J Ayub Coll Abbottabad* 2007; 19: 26-28.

55. Memon IA, Jamal A, Arif F, Murtaza G. Causes of non-polio AFP in children residing in the province of Sindh. *Medical Channel Pediatrics* 2010:16 (3).

56. D'Souza RM, Kennett M, Antony J. Surveillance of acute flaccid paralysis in Australia: Australian Paediatric Surveillance Unit, 1995–97. *J. Paediatr. Child Health* 1999; **35**: 536-40.

57. Chan LG, Parashar UD, Lye MS. Deaths of children during an outbreak of hand, foot and mouth disease in Sarawak, Malaysia: clinical and pathological characteristics of the disease. *Clin. Infect Dis* 2000; **31**: 678-83.

58. Oostvogel P, Spaendonck M, Hirasing R. Surveillance of acute flaccid paralysis in the Netherlands,1992-94. *Bull World Health Organ* 1998; **76**(1): 55.

59. Tal-hatu KH, Omotade TT. Acute flaccid paralysis: a five-year review of cases managed by physiotherapy at the University College Hospital, Ibadan. *Afr J Health Sci* 2006; **13**(1-2): 28-32.

DS (Child) H J 2018; 34(1) : 108-111

ORIGINAL ARTICLE

Pattern of Spirometry among Asthmatic Patients of Paediatric Age Group Who Attend the Outpatient's Clinic with Respiratory Symptoms

Md. Mosharaf Hossain[1], Md. Shakibur Rahman[2], Mohammed Rizwanul Ahsan[3], Md. Kamrunzzaman Kamrul[1]

Abstract

Background: *Asthmatic patients typically show obstructive patterns on spirometry with positive bronchodilator responses. Spirometry is a valuable tool for the diagnosis, step care management and follow-up. Asthma is a heterogeneous disease, usually characterized by chronic airway inflammation.*

Objective: *To find out the pattern of spirometry among asthmatic patients of paediatric age group who attend the outpatient's clinic with respiratory symptoms.*

Methods: *A cross-sectional study was conducted among 60 asthmatic patients who attended the outpatient clinic with the following respiratory symptoms (cough, wheezes, chest tightness, and shortness of breath) at Dhaka Shishu (Children) Hospital, Sher-e-Bangla Nagar, Dhaka during November 2017 to September 2018. All the participants were known asthmatic for at least one year before their attendance.*

Results: *Twenty two (36.7%) patients were found restrictive, 20 (33.3%) were mixed, 7 (11.7%) were obstructive and 11 (18.3%) were normal. In this study the reversibility tests were positive in significant proportions of the participants, including those who presented with non-obstructive patterns (*$p < 0.001$*).*

Conclusion: *Non-obstructive patterns of spirometry findings were predominant among paediatric patient suffering from asthma where reversibility test also positive in significant proportion.*

Key words: *Spirometry, bronchial asthma.*

Introduction

Asthmatic patients typically show obstructive patterns on spirometry with positive bronchodilator responses; however, other spirometric patterns were also reported.[1] Bronchial asthma is a major health problem that affects more than 300 million people worldwide. Its diagnosis depends on the clinical presentation and the objective evidence of a reversible airflow obstruction or airway hyper-responsiveness. Spirometry is a valuable tool for the diagnosis and follow-up of asthma excerbation.[2,3] Asthma is a heterogeneous disease, usually characterized by chronic airway inflammation.[4] It is defined by the history of respiratory symptoms such as wheezing, breathlessness, chest tightness, and coughing, which vary over time and in intensity, along with variable

1. Assistant Professor, Department of Pediatric Respiratory Medicine (Pulmonology), Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital, Dhaka.
2. FCPS Trainee, Department of Pediatric Respiratory Medicine (Pulmonology), Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital, Dhaka.
3. Assistant Professor, Bangladesh Institute of Child Health (BICH) & Resident, Dhaka Shishu (Children) Hospital, Dhaka.

Correspondence to: Dr. Md. Mosharaf Hossain, Assistant Professor, Department of Pediatric Respiratory Medicine (Pulmonology), Dhaka Shishu (Children) Hospital, Dhaka. Cell: 01710973500, E-mail: mosharafhossain1966@yahoo.com
Received: 10 October 2018; **Accepted**: 6 November 2018

expiratory airflow.[5] Given the rising incidence of non-communicable diseases (NCDs) globally, especially bronchial asthma, there is the need to reduce the associated morbidity and mortality by adopting an objective means of diagnosis and monitoring.[6]

Materials and Methods

A cross-sectional study was conducted among 60 asthmatic patients who attended the outpatient clinic with the following respiratory symptoms (cough, wheezes, chest tightness, and shortness of breath) in Dhaka Shishu (Children) Hospital, Sher-e-Bangla Nagar, Dhaka during November 2017 to September 2018. All the participants were known asthmatic for at least one year before their attendance.

A portable All-flow spirometer was used for lung function measurements. All the measurements were performed according to the guidelines of the American Thoracic Society (ATS). Reversibility test was done by bronchodilator (sulbutamol) nebulization and reversibility test was considered positive when post bronchodilatation FEV_1 increasing 12% or 200 ml from previous FEV_1. Written consents were obtained from the patients before starting the spirometry measurements.

Data were analyzed using the Statistical Package for the Social Sciences (SPSS) Inc. Chicago, IL, USA) ver-sion 23. Mean and standard deviations were used to describe the spirometric values. Categorical variables were compared using the chi-square test. Statistical significance was accepted when the *p*-value <0.05.

Results

Mean age was 9.1±2.8 years, 39 (65.0%) patients were male and 23 (38.3%) were with normal BMI (Table I).

Table-I
Demographic characteristics of the study population

Demographic characteristics	Number of patients	Percentage
Age (years)		
≤5	10	16.7
6-10	38	63.3
>10	12	20.0
Mean±SD	9.1±2.8	
Range (min-max)	4-15	
Sex		
Male	39	65.0
Female	21	35.0
BMI (Kg/M^2)		
Low (<18.5)	8	13.3
Normal (18.2- 24.9)	23	38.3
Over weight (25.0-29.9)	19	31.7
Obese (≥30)	10	16.7

Mean FVC was 2.6±0.9 L, FEV1 was 2.1±0.8 L, FEV1/FVC was 79.9±12.3 percent, FEF 25 was 3.6±1.6, FEF 50 was 2.4±1.3, FEF 75 was 1.2±0.8, FEF 25-75 was 2.2±1.2 and PEFR was 4.1±1.6 (Table II).

Table II
Pulmonary function test of the study population

Pulmonary function test	Mean±SD
FVC (L)	2.6±0.9
FEV1 (L)	2.1±0.8
FEV1/ FVC (%)	79.9±12.3
FEF 25	3.6±1.6
FEF 50	2.4±1.3
FEF 75	1.2±0.8
FEF 25-75	2.2±1.2
PEFR	4.1±1.6

Twenty two (36.7%) patients was found restric-tive, 20 (33.3%) was mixed, 7 (11.7%) was obstructive and 11 (18.3%) was normal spirometry (Table-III).

Table III
Pattern of spirometry of the study population

Pattern of spirometry	Noumber of patients	Percentage
Normal	11	18.3
Mixed	20	33.3
Obstructive	7	11.7
Restric-tive	22	36.7

Reversibility tests were positive in significant proportions of the participants, including those who presented with non-obstructive patterns ($p < 0.001$) (Table-IV).

Table IV
Association between variables of spirometry with Pulmonary function test

Pulmonary function test	Total	Pattern of spirometry								p*
		Normal		Restrictive		Obstructive		Mixed		
		n	%	n	%	n	%	n	%	
FVC (ml)										
Abnormal	**48**	3	27.3	19	86.4	7	100.0	19	95.0	0.001[s]
Normal	**12**	8	72.7	3	13.6	0	0.0	1	5.0	
FEV1 (ml)										
Abnormal	**47**	4	36.4	17	77.3	7	100.0	19	95.0	0.001[s]
Normal	**13**	7	63.6	5	22.7	0	0.0	1	5.0	
FEF25-75										
Abnormal	**45**	1	9.1	18	81.8	7	100.0	19	95.0	0.001[s]
Normal	**15**	10	90.9	4	18.2	0	0.0	1	5.0	

* Chi-square test; s= significant

Discussion

In this study it was observed that mean age was 9.1±2.8 years, 39(65.0%) patients were male and 23(38.3%) were normal BMI. In study of Merghani,[1] observed that the mean body mass index was 28.6±6.6 kg/m^2. Salviano et al.[7] study reported average age was 11.5±3.0 years, with a predominance of males (66.7%). Maheswari et al[8] showed that affected patients were more often males 75.4% than females 24.5%. In this series it was observed that mean FVC was 2.6±0.9 L, FEV_1 was 2.1±0.8 L, FEV_1/FVC was 79.9±12.3 percent, FEF 25 was 3.6±1.6, FEF 50 was 2.4±1.3, FEF 75 was 1.2±0.8, FEF 25-75 was 2.2±1.2 and PEFR was 4.1±1.6. Similar observation was found different studies, in study of Merghani showed that the obstruction could be within the middle and the small airways, as indicated by the low values of the forced expir-atory flows (FEF 25, FEF 50, FEF 75 and FEF 25-75).[1] Salviano et al.[7] study observed that the mean FVC was 2.8 ml, FEV_1 was 2.26 ml and FEV_1/ FVC was 80.1 percent.

In this present study it was observed that 7 (11.7%) was obstructive and 53 (88.3%) was non-obstructive spirometry. Similar observation was found Merghani,[1] study observed that patterns of spirometry were obstructive (12%), normal (21%), restric-tive (36%) and mixed (32%). Salviano et al[7] showed out of 90 children 55(61.1%) had obstructive and 35(38.9%) had non-obstructive pattern.

In this study it was observed that the reversibility tests were positive in significant proportions of the participants, including those who presented with non-obstructive patterns ($p < 0.001$). Similar result was found in Merghani.[1] One of the possible explanations for the restrictive pattern is the high closing capacity in asthmatic patients that increases air trapping and decreases the overall volume of air that ventilates the lungs; thus reducing FEV1 and FVC values.[9] Obesity could be another explanation; however, the association in this study was statistically insignificant.

Conclusion

Non-obstructive patterns of spirometry findings were predominant among paediatric patient suffering from asthma where reversibility test was also positive in significant proportion.

References

1. Merghani TH. Patterns of spirometry in asthmatic patients presenting with respiratory symptoms. *Int J Med Sci Public Health* 2017; **6**: 337-40.
2. Global Initiative for Asthma (GINA). Global strategy for asthma management and prevention (2016 updates); available on: http:// ginasthma.org /2016-gina-report-global-strategy-for-asthma management-and-prevention/ Accessed 3/8/2016.
3. British Thoracic Society/ COPD Consortium. Spirometry in practice: a practical guide to using spirometry in primary care. 2nd ed. April 2005.
4. Chang AB, Robertson CF, van Asperen PP, et al. A multi-centre study on chronic cough in children: burden and etiologies based on astandardized management pathway. *Chest* 2012; **142**(4): 943-50.
5. Marchant JM, Newcombe PA, Juniper EF, et al. What is the burden of chronic cough for families? *Chest*. 2008; **134**(2): 303-09.

6. Ayuk AC, Uwaezuoke SN, Ndukwu CI, Ndu IK, Iloh KK, Okoli CV. Spirometry in Asthma Care: A Review of the Trends and Challenges in Pediatric Practice. *Pediatrics,* 2017; **11**: 1-6.

7. Salviano LDDS, Taglia-Ferre KD, Lisboa S, Costa ACCD, Campos HDS, March MFP. Association between fraction of exhaled nitric oxide and spirometry data and clinical control of asthma in children and adolescents. *Rev Paul Pediatr* 2018; **36** (1): 17-24.

8. Maheswari K, Wadhwa L. Clinical profile and causes of mesenteric lymphadenitis in children. *Int J Contemp Pediatr* 2016; **3**(2): 550-52.

9. Keddissi JI, Elya MK, Farooq SU, Youness HA, Jones KR, Awab A, et al. Bronchial Responsiveness in Patients with Restrictive Spirometry. BioMed Research International 2013; 2013, Article ID 498205.

DS (Child) H J 2018; 34(1) : 112-118

REVIEW ARTICLE

Congenital Heart Block: A Review

Mohammad Abdullah Al Mamun[1], Rezoana Rima[2], Abdul Jabbar[3], Manzoor Hussain[4]

Abstract

Congenital heart block is a rare disorder with an incidence of 1 in 15,000-22,000 live births. It is frequently associated with underlying structural congenital heart disease (CHD). Congenital heart block associated with neonatal lupus is considered a form of passively acquired autoimmune disease in which maternal autoantibodies to the intracellular ribonucleoproteins Ro (SS-A) and La (SS-B), cross the placenta and injure the previously normal fetal heart. With the advancement of fetal echocardiography, a high proportion of cases of congenital heart block are now identified in utero. The finding may be incidental or investigations may have been performed because of concerns regarding fetal development. It may be associated with high mortality and morbidity specially those associated with structural heart disease. As this is an uncommon disorder and associated with adverse outcome therefore requires a high index of suspicion for early diagnosis.

Keywords: *Congenital, heart block.*

Introduction

Presence of conduction system disease of any form, which is diagnosed on or before 28 days of life is considered as congenital heart block.[1] Increasing numbers of cases of congenital heart block are being diagnosed between 18 and 24 weeks of gestation due to increasing prenatal care and use of ultrasound technology in pregnancy. The incidence of congenital heart block has been estimated from several studies to be about 1 in 15,000-22,000 live births.[1-3] Congenital heart block may occur with or without associated structural heart disease. Approximately one third of patients with cardiac conduction defects has congenital heart defect.[4,5] In utero, diagnosis of congenital heart block is associated with structural heart disease in approximately one half of the cases.[3] In isolated congenital heart block, maternal autoantibodies to SS-A/Ro and/or SS-B/La proteins cross the placenta and may lead to permanent destruction of the fetal atrioventricular (AV) conduction system and produces heart block.[6]

In congenital heart block there may be delayed conduction of impulse from atrium to AV node or His-Purkinje system evidenced by a prolonged PR interval called First-degree heart block. Second-degree heart block is characterized by intermittent failure of conduction to the ventricle. It is classified further into Mobitz type I or II block. Mobitz type I or Wenckebach block is due to blockage in the AV node. Its characteristic appearance on electro-cardiography (ECG) is gradual prolongation of the PR interval with eventual failure of conduction and dropped beat. Mobitz

1. Associate Professor, Division of Neonatal Cardiology, Department of Pediatric Cardiology, Bangladesh Institute of Child Health and Dhaka Shishu (Children) Hospital.
2. Associate Professor and Interventional Cardiologist, Department of Pediatric Cardiology, Bangladesh Institute of Child Health and Dhaka Shishu (Children) Hospital.
3. Registrar, Department of Pediatric Cardiology, Dhaka Shishu (Children) Hospital.
4. Professor and Head, Division Pediatric Medicine and Department of Pediatric Cardiology, Bangladesh Institute of Child Health and Dhaka Shishu (Children) Hospital.

Correspondence to: Dr. Mohammad Abdullah Al Mamun, Associate Professor, Division of Neonatal Cardiology, Department of Pediatric Cardiology, Bangladesh Institute of Child Health and Dhaka Shishu (Children) Hospital, Dhaka. Cell: 01913475529, E-mail: mamundsh@gmail.com

Received: 10 May 2018; **Accepted**: 2 June 2018

type II block, is due to blockage in the distal conducting system. There is no lengthening of the PR interval prior to the blocked beat. An abnormality of repolarization of the ventricle called Long QT syndrome can present with an extremely long QT interval and associated 2:1 AV block due to ventricular refractoriness. Complete heart block (CHB) is characterized by no conduction from the atrium to the ventricle. The atrial rate is higher than the ventricular rate, and P waves have no relationship to the QRS complexes (Fig. 1).[7]

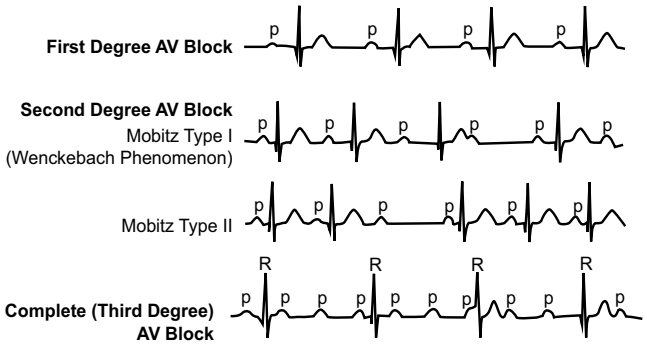

Fig.-1 *In First-degree AV block, the PR interval is prolonged, but all the atrial impulses are conducted to the ventricle. In Second-degree AV block, not every atrial impulse is conducted to the ventricle. In Mobitz type I, classically the PR interval increases progressively until a P wave is not conducted. In Mobitz type II, there is no progressive conduction delay and subsequent shortening of the PR interval after a blocked beat. In Third-degree AV block or complete heart block, P-P interval normal and constant, QRS complex normal but there is atrioventricular dissociation and atrial rate is higher than ventricular rate.*

Congenital heart block associated with structural heart disease

Congenital heart block is frequently associated with underlying structural CHD. The commonest forms of CHD associated with heart block include left atrial isomerism. Atrioventricular septal defect and congenitally corrected transposition of the great arteries (ccTGA) have also been identified as causes of congenital heart block. Congenital heart block with CHD may be responsible for more than half of all prenatally diagnosed cases of heart block.[3]

Congenital heart block associated with maternal autoantibodies

Autoantibody-associated congenital heart block is a passively acquired autoimmune condition in which maternal autoantibodies are thought to initiate conduction disturbances in the developing fetal heart. It is believed to be the consequence of an inflammatory response that results in tissue injury, fibrosis, and scarring of the conduction system. The transplacental entry of autoantibodies into the fetal circulation are believed to be responsible for triggering this response.[8] Using the recently developed technique of line immunoassay, it has been possible to detect SS-A/Ro or SS-B/La antibodies in the sera of 95% of the mothers of fetuses or newborns presenting with AV block.[9] The risk for congenital heart block in an anti-Ro positive pregnancy is 1% to 2%, and a reported recurrence rate of 12% to 20%.[10,11] Complete AV block is the major manifestation of autoantibody-associated congenital heart block, other cardiac abnormalities are increasingly being recognized like transient first-degree AV block has been shown to occur in up to 30% of fetuses of mothers with anti-SS-A/Ro antibodies and presence of sinus bradycardia and prolongation of the QTc interval have also been reported.[12-14] Clinical signs most commonly develop during 18 to 24 weeks of pregnancy. Although autoantibody-associated congenital heart block may initially be detected as a first- or second-degree AV block, most of the affected pregnancies will present with fetal bradycardia in third-degree (complete) AV block, and ventricular rates typically are between 50 and 70 beats per minute.[15,16]

Pathophysiology

Structural heart disease associated congenital heart block: In association with structural heart disease, heart block is thought to result from an anatomical discontinuity of the electrical conduction system, either due to an initial lack of fusion between AV nodal tissue and the His bundle or due to a secondary interruption of the AV conduction axis.[17]

Autoantibody-associated congenital heart block: Hallmarks of autoantibody-associated congenital heart block are the presence of immune complex deposits, inflammation, calcification, and fibrosis in the fetal heart and a block in signal conduction at the AV node in an otherwise structurally normal heart.[10] Development of congenital heart block is closely associated with the presence of maternal autoantibodies that are most commonly found in the rheumatic autoimmune diseases Sjögren's syndrome (SS) and systemic lupus erythematosus (SLE).[18] However, congenital heart block appears to be more specifically linked to the presence of maternal anti-Ro/SS-A and anti-La/SS-B autoantibodies rather than to maternal diagnosis, as the mother of an affected child may be asymptomatic.[19,20] This finding was further confirmed recently in a population based study

in Sweden, where the serum of 95% and 63% of autoantibody positive women who had a child with heart block displayed reactivity toward the Ro52 and Ro60 proteins, respectively.[21] In contrast to the association of anti-Ro antibodies to congenital heart block, the association of anti-La antibodies to congenital heart block is still a matter of debate.[22,23]

Neonatal lupus associated congenital heart block: The mechanism of causation of neonatal lupus associated congenital heart block is not completely understood but evidence points to the fetus beginning life with a normal cardiac structure and conduction system. At approximately 12 weeks of gestation, maternal IgG antibodies against Ro and La intracellular ribonuclear proteins are actively transported across the placenta and are thought to bind specific cells of the fetal conduction system. This may result in a cycle of inflammation, later scarring and fibrosis.[1] Neonatal lupus is usually diagnosed in the presence of a slow heart rate discovered in a fetus or newborn in the absence of associated structural cardiac abnormalities. Other manifestations of neonatal lupus may include the presence of skin rashes, liver abnormalities determined biochemically and abnormalities in the cellular elements of the blood including various cytopenias.[24] Maternal serum testing subsequently reveals antibodies to Ro and/or La. In utero, the peak onset of the diagnosis of bradycardia is between 18 and 24 weeks of gestation, corresponding to the window of opportunity about six weeks after effective placental transport of maternal IgG antibodies begin. The degree of heart block may vary from first degree to third degree block, but most cases diagnosed in utero present with at least second degree or more advanced block. The occurrence rate of neonatal lupus has been estimated at approximately 2 to 3% in all pregnancies born to women with anti-Ro or anti-La antibodies. The recurrence rate in a mother with antibodies who has a previous child who was affected, is approximately 18%.[25]

Clinical presentation

With the advent and development of fetal echocardiography, a high proportion of cases of autoimmune-mediated congenital heart block are now identified in utero (in populations where routine fetal echocardiography is performed). The finding may be an incidental finding, or alternatively, investigations may have been performed because of concerns regarding fetal development.[26] Postnatally, neonates and young children present in a wide variety of different ways. Some are asymptomatic with bradycardia and detected only during routine screening. Others may present with nonspecific symptoms, such as poor growth, abnormal tiredness, sleep disturbance, or frequent nightmares. Syncope, heart failure, or sudden death may be the first manifestation of congenital AV block in the absence of previous symptoms and signs of cardiovascular disease.[19]

Investigations

Fetal echocardiography: The majority of cases of congenital heart block diagnosed in utero are detected by either auscultation or routine obstetrical ultrasound in low risk pregnancies. Despite recent developments in the field of electro-and magnetocardiography, fetal echocardiography remains the gold standard method for diagnosing congenital heart block.[27] The diagnosis is made by establishing atrioventricular dissociation by using M-mode or Doppler echocardiographic techniques. Fetal LVOT Doppler can measure mechanical PR interval by detecting long pause between the onset of the atrial contraction and onset of ejection time (PR interval). It can detect 1st degree heart block with the addition of profound sinus bradycardia, Wenckebach Mobitz Type I, a type of 2nd degree heart block. Spectral Doppler may show a rapid atrial rate than ventricular rate with atria and ventricular dissociation in 3rd degree heart block.[1]

Electrocardiogram: Complete AV block is by far the most common finding in children with AV conduction impairment. Sinus bradycardia, 1st degree AV block with prolong PR interval, 2nd degree AV block which is characterized by gradual prolongation of the PR interval with eventual failure of conduction and dropped beat, Mobitz type II block where there is no lengthening of the PR interval prior to the blocked beat and prolong QTc interval (corrected QT interval >450 ms) can be detected by ECG.[28,29] 24-hour electrocardiographic Holter monitoring is helpful as part of assessment of patients with congenital heart bock.

Echocardiography: Echocardiography is an important investigation in the postnatal population. It allows intracardiac structural malformations to be identified and provides an assessment of ventricular function.[30] 2D-directed M-mode echocardiogram may detect a very poorly contractile, dilated ventricle with barely thickened ventricular wall during systole.

Others: Once heart block has been diagnosed, the mother should be screened for evidence of connective tissue disease. Immunoassays for SS-A/Ro or SS-B/La antibodies should be performed.

Natural history of patients with congenital AV block

Spontaneous recovery from congenital AV block is very rare, though it has been reported in exceptional cases.[31] The estimated overall mortality of nonpaced patients with isolated complete CHB is 8%-16% in infants and 4%-8% in children and adults. The high mortality observed in neonates appears to occur as a consequence of the complications of prematurity and bradycardia owing to the delayed initiation of pacing therapy. Untreated, congenital, antibody-mediated AV block is associated with a fetal and neonatal mortality ranging between 14% and 34%.[32] The morbidity and mortality associated with CHB remains high even as the child grows and becomes an adult.[29] There is a high mortality rate, particularly in fetuses diagnosed in utero with hydrops, and it is approximately 20%. Bradycardia alone is not always the full extent of disease. Relatively high incidence of the development of late cardiomyopathy leading to heart failure, death or transplantation despite successful pacemaker implantation.[33]

Treatment

Prenatal management: Treatment for identified congenital heart block in utero may be effective if it can reduce a generalized inflammatory insult and lower the titer of maternal autoantibodies, several prenatal therapeutic protocols have been utilized. These include the use of adrenocorticosteroids, which are not metabolized by the placenta, principally dexamethasone. Some researchers have also attempted plasmapheresis and the use of maternal alpha adrenergic agents.Use of these significantly lower fetal mortality in a recent study.[32]

If the heart block is already third degree that means CHB and has been present for more than three weeks, an attempt at reversing this is futile and therefore serial echocardiographic and obstetrical follow-up should require without any therapy. If, however, the third degree or CHB has been recently diagnosed, a therapeutic course of dexamethasone 4 mg orally once a day for a period of six weeks may be tried. If there has been no change in fetal status, tapering the course and discontinuation should be done. On the other hand, if the fetus' conduction system disease has improved to second degree block or better, then dexamethasone should be continued until delivery and subsequently taper in the mother.[1]

If the fetus presents with alternating second and third degree block, dexamethasone at 4 mg orally daily for a six-week period of time should be used. If the conduction system disease progresses to third degree block then drug should be stopped. But if there has been improvement to second degree or better, steroids should be continued until delivery and taper thereafter.[1]

If the fetus is discovered to have only second degree or a simply prolonged mechanical PR interval (first degree block), then dexamethasone 4 mg orally daily should be given until delivery and taper the dose after that. On the other hand, if this early block progresses to permanent third degree block and present for six weeks or longer steroid should be stopped.[1,34]

Maternal use of dexamethasone is not without risks. There is the potential for harm to both mother and baby as a result of steroid use, in particular fetal neurological development may be impaired.[35] Glucocorticoid associated risks of increased infection, loss of bone density, diabetes, hypertension and cataracts may occur. The fetal risks of maternal steroids include oligohydramnios, intrauterine growth retardation and adrenal suppression. Percutaneous in utero pacing has been investigated as a method for providing heart rate support.

Postnatal management: Both the North American and European guidelines recommend implantation of a pacemaker in children with third degree or CHB and any of the following symptoms: syncope, congestive heart failure, or chronotropic incompetence with the limitation of physical activity etc. and patients with more subtle symptoms such as tiredness, poor growth, frequent nightmares, or naps. If clinically bradycardia is felt to be the most likely cause of the symptoms, then the recommendation is usually to advise pacing.[36] An observational study showed that cardiac pacing is beneficial in asymptomatic patients presenting with echocardiographically apparent left ventricular (LV) dilatation or dysfunction.[37] Patients who had a heart rate <50 beats/min or pauses >3 seconds on Holter monitoring had an increased incidence of syncope and sudden death.[38] There is good evidence that QT prolongation is a risk factor for syncope or sudden death in patients with congenital heart block.[39] Practice is to implant a pacemaker in asymptomatic patients when heart rate is found to be <55 beats/min in neonates or <50 beats/min in children, adolescents, and adults.[1]

Outcome of paced patients: After the implantation of a pacemaker, it was often assumed that children presenting with third degree or CHB would go on to enjoy an average quality of life. However, various studies have demonstrated that some patients are prone to develop severe systolic dysfunction despite the early institution of cardiac pacing.[40,41] Despite early institution of cardiac pacing, some infants with CHB developed cardiomyopathy.[33]

Prognosis

If the heart block is diagnosed as bradycardia during the fetal period, there is a very high rate of fetal and neonatal death. Prenatal risk factors for mortality depend on the presence of structural heart disease and a heart rate less than a critical value, frequently quoted as 55 bpm. The presence of hydrops fetalis or other signs of physiologic disturbance in cardiac function, are very poor prognostic signs. In severe cases, there has been as high as an 85% mortality rate in the neonatal period. If the congenital heart block is first diagnosed in the newborn period, the prognosis is somewhat better. Once again, the presence or absence of underlying structural heart disease often determines the outcome. The survival rate in newborns with congenital heart block and no structural heart disease is about 85%. Many of these patients require pacemaker implantation. If the congenital heart block first presents beyond the newborn period, patients are unlikely to have severe structural heart disease, and the survival rate is much higher than 85%. They require pacemaker implantation as well as treatment for any underlying structural heart disease. In later childhood or adulthood patients are unlikely to have structural heart disease and they tend to have a good prognosis after pacemaker implantation.[1]

Conclusion

Congenital heart block is associated with higher mortality and morbidity specially those having structural heart disease. With the advancement of fetal echocardiogram now high proportion of cases are identified in utero. Outcome can be improved with early diagnosis and aggressive therapy therefore requires a high index of suspicion.

References

1. Friedman DM, Duncanson LJ, Glickstein J, BuyonJP. A review of congenital heart block. *Images Paediatr Cardiol* 2003; **5**(3): 36-48.
2. Yater W. Congenital heart block: review of the literature. *Am J Dis Child* 1929; **38**: 112-136.
3. Kertesz NJ, Fenrich AL, Friedman RA. Congenital complete atrioventricular block. *Texas Heart Inst J* 1997; **24**: 301-307.
4. Ross BA. Atrioventricular block. In: Garson A Jr, Bricker JT, McNamara DG, editors. The science and practice of pediatric cardiology. Philadelphia, PA: Lea and Febiger; 1990. p 1803.
5. Jaeggi ET, Hornberger LK, Smallhorn JF, FouronJC. Prenatal diagnosis of complete atrioventricular block associated with structural heart disease: combined experience of two tertiary care centers and review of the literature. *Ultrasound Obstet Gynecol* 2005; **26**: 16-21.
6. Waltuck J, Buyon JP. Autoantibody-associated congenital heart block: Outcome in mothers and children. *Ann Intern Med* 1994; **120**: 544-51.
7. Dubin AM. Arrhythmias in the Newborn. *NeoReviews* 2000;**1**:e146-e151.
8. Taylor PV, Scott JS, Gerlis LM, Esscher E, Scott O. Maternal antibodies against fetal cardiac antigens in congenital complete heart block. *N Engl J Med* 1986; **315**: 667-72.
9. Costedoat-Chalumeau N, Georgin-Lavialle S, Amoura Z, Piette JC. Anti-SSA/Ro and anti-SSB/La antibody-mediated congenital heart block. *Lupus* 2005; **14**: 660-64.
10. Ambrosi A, Salomonsson S, Eliasson H, Zeffer E, Skog A, Dzikaite V et al. Development of heart block in children of SSA/SSBautoantibody-positive women is associated with maternal age anddisplays a season-of-birth pattern. *Ann Rheum Dis* 2012, **71**: 334-40.
11. Llanos C, Izmirly PM, Katholi M, Clancy RM, Friedman DM, Kim MY et al. Recurrence rates of cardiac manifestations associated with neonatal lupusand maternal/fetal risk factors. *Arthritis Rheum* 2009; **60**: 3091-97.
12. Sonesson SE, Salomonsson S, Jacobsson LA, Bremme K, Wahren-Herlenius M: Signs of fi rst-degree heart block occur in one-third of fetuses of pregnantwomen with anti-SSA/Ro 52-kd antibodies. *Arthritis Rheum* 2004; **50**: 1253-61.
13. Brucato A, Frassi M, Franceschini F, CimazR, Faden D, Pisoni MP, et al. Risk of congenital complete heart block in newborns of mothers with anti-Ro/SSA antibodies detected by counter immune electrophoresis: a prospective study of 100 women. *Arthritis Rheum* 2001; **44**: 1832-35.
14. Gerosa M, Cimaz R, Stramba-Badiale M, Goulene K, Meregalli E, Trespidi L, et al. Electrocardiographic

abnormalities in infants bornfrom mothers with autoimmune diseases-a multicentre prospective study. *Rheumatology (Oxford)* 2007; **46**: 1285-89.

15. Eronen M, Siren MK, Ekblad H, Tikanoja T, Julkunen H, Paavilainen T: Shortand long-term outcome of children with congenital complete heart block diagnosed in utero or as a newborn. *Pediatrics* 2000; **106** (1 Pt 1): 86-91.
16. Ambrosi A, Wahren-Herlenius M. Congenital heart block: evidence for a pathogenic role of maternal autoantibodies. *Arthritis Research & Therapy* 2012; **14**: 208.
17. Ho SY, Fagg N, Anderson RH, Cook A, Allan L. Dispositionof the atrioventricular conduction tissues in the heart with isomerism of the atrial appendages: its relation to congenital complete heart block. *J Am Coll Cardiol* 1992; **20**: 904-10.
18. Taylor PV, Taylor KF, Norman A, Griffiths S, Scott JS: Prevalence of maternal Ro (SS-A) and La (SS-B) autoantibodies in relation to congenital heart block. *Br J Rheumatol* 1988, **27**: 128-32.
19. Jaeggi ET, Hamilton RM, Silverman ED, Zamora SA, Hornberger LK: Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block. A single institution's experience of 30 years. *J Am Coll Cardiol* 2002, **39**: 130-37.
20. Llanos C, Izmirly PM, Katholi M, Clancy RM, Friedman DM, Kim MY, Buyon JP: Recurrence rates of cardiac manifestations associated with neonatal lupus and maternal/fetal risk factors. *Arthritis Rheum* 2009; **60**: 3091-97.
21. Salomonsson S, Dzikaite V, Zeffer E, Eliasson H, Ambrosi A, Bergman G, et al. A population-based investigation of the autoantibody profi le in mothers of children with atrioventricular block. *Scand J Immunol* 2011; 74: 511-17.
22. Jaeggi E, Laskin C, Hamilton R, Kingdom J, Silverman E. The importance of the level of maternal anti-Ro/SSA antibodies as a prognostic marker of the development of cardiac neonatal lupus erythematosus a prospective study of 186 antibody-exposed fetuses and infants. *J Am Coll Cardiol* 2010; **55**: 2778-84.
23. Gordon P, Khamashta MA, Rosenthal E, Simpson JM, Sharland G, Brucato A, et al. Anti-52 kDa Ro, anti-60 kDa Ro, and anti-La antibody profi les in neonatal lupus. *J Rheumatol* 2004; **31**: 2480-87.
24. Friedman DM, Rupel A, Glickstein J, Buyon JP. Congenital heart block in neonatal lupus: The pediatric cardiologist's perspective. *Indian J Pediatr* 2002; **69**: 517-22.
25. Buyon JP, Heibert R, Copel J. Autoimmune-associated congenital heart block: Mortality, morbidity, and recurrence rates obtained from a national neonatal lupus registry. *J Am Coll Cardiol* 1998; **31**: 1658-66.
26. Fouron JC, Proulx F, Miro J, Gosselin J. Doppler and M-mode ultrasonography to time fetal atrial and ventricular contractions. *Obstet Gynecol* 2000; **96**: 732-36.
27. Andelfinger G, Fouron JC, Sonesson SE, Proulx F. Reference values for time intervals between atrial and ventricular contractions of the fetal heart measured by two Doppler techniques. *Am J Cardiol* 2001;**88**:1433-36.
28. Askanase AD, Friedman DM, Copel J, et al. Spectrum and progression of conduction abnormalities in infants born to mothers with anti-SSA/Ro-SSB/La antibodies. *Lupus* 2002; **11**: 145-51.
29. Michaelsson M, JonzonA, Riesenfeld T. Isolated congenital complete atrioven-tricular block in adult life: a prospective study. *Circulation* 1995; **92**: 42-49.
30. Beaufort-Krol G, Schasfoort-Van Leeuwen MJ, Stienstra Y, Bink-Boelkens M. Longitudinal echocardiographic follow-up in children with congenital complete atrioventricular block. *Pacing Clin Electrophysiol* 2007; **30**: 1339-43.
31. De Raet J, Rega F, Meyns B. Late recovery of atrioventricular conduction after pacemaker implantation for complete heart block in congenital heart disease: fact or fluke? *Acta Chir Belg* 2010; **110**: 323-27.
32. Jaeggi ET, Fouron JC, Silverman ED, Ryan G, Smallhorn J, Hornberger LK. Transplacental fetal treatment improves the outcome of prenatally diagnosed complete atrioventricular block without structural heart disease. *Circulation* 2004; **110**: 1542-48.
33. Moak JP, Barron KS, MD, Hougen TJ, Wiles HB, Balaji S, Sreeram N, et al. Congenital heart block: development of late-onset cardiomyopathy, a previously underappreciated sequela. *J Am Coll Cardiol* 2001; **37**: 238-42.
34. Glickstein JS, Buyon JP, Friedman D. Pulsed Doppler echocardiographic assessment of the fetal PR interval. *Am J Cardiology* 2000; **86**: 236-239.
35. Pisoni CN, Brucato A, Ruffatti A. Failure of intravenous immunoglobulin to prevent congenital heart block: findings of a multicenter, prospective,

observa- tional study. *Arthritis Rheum* 2010; **62**: 1147-52.

36. Bordachar P, Zacharay W, Whinnett Z, Ploux S, Labrousse L, Haissaguerre M, Thambo JB. Pathophysiology, clinicalcourse, and management of congenital complete atrioventricular block. *Heart Rhythm* 2013; **10**: 760-66.
37. Beaufort-Krol G, Schasfoort-Van Leeuwen MJ, Stienstra Y, Bink-Boelkens M. Longitudinal echocardiographic follow-up in children with congenital complete atrioventricular block. Pacing Clin Electrophysiol 2007; **30**: 1339-43.
38. Dewey RC, Capeless MA, Levy AM. Use of ambulatory electrocardiographic monitoring to identify high-risk patients with congenital complete heart block. *N Engl J Med* 1987; **316**: 835-39.
39. Berul CI, Cecchin F. American Heart Association American College of Cardiology. Indications and techniques of pediatric cardiac pacing. *Expert Rev Cardiovasc Ther* 2003; **1**: 165-76.
40. Rozmus G, Daubert JP, Huang DT, Rosero S, Hall B, Francis C. Venous thrombosis and stenosis after implantation of pacemakers and defibrillators. J Interv Card Electrophysiol 2005; **13**: 9-19.
41. Bar-Cohen Y, Berul CI, Alexander ME, et al. Age, size, and lead factors alone do not predict venous obstruction in children and young adults with transvenous lead systems. *J Cardiovasc Electrophysiol* 2006; **17**: 754-59.

DS (Child) H J 2018; 34(1) : 119-122

REVIEW ARTICLE

Diagnosis and Management of Hirschsprung Associated Enterocolitis

Md Ayub Ali[1], Md Samiul Hasan[2]

Abstract

The diagnosis of Hirschsprung-associated enterocolitis (HAEC) is often difficult to establish because the clinical signs and symptoms are nonspecific. An evidence-based rational approach to the diagnosis and management of HAEC is indispensable to reduce morbidity & mortality and to improve the quality of life of patients with HD. Etiologies of HAEC include dysbiosis of the intestinal microorganism, impaired mucosal barrier function, altered innate immune responses, and bacterial translocation. The clinical suspicion and severity of HAEC have been categorized into three grades based on history, physical examination, and imaging studies. The goal of this approach is to produce a standard, clinically relevant, and convenient to use a system that can be universally adopted to allow a consistent approach to the diagnosis and treatment of HAEC. The severity of the clinical presentation directs the treatment of HAEC. Close monitoring is essential to detect symptoms progression to a higher grade of disease. Additional evaluation is required in children with recurrent HAEC to determine whether a cause can be identified.

Keywords: *Hirschsprung disease, hirschsprung associated enterocolitis.*

Background

Since its first description by Härold Hirschsprung, a significant improvement has been noted in the management of Hirschsprung disease (HD) over the last few decades.[1] Despite this fact, Hirschsprung-associated enterocolitis (HAEC) is still the common and leading cause of morbidity and mortality of patients with HD. It is an inflammatory disorder of the bowel that was first described by Swenson in 1956.[1,2] Patients with Hirschsprung disease (HD) are in lifetime risk for HAEC. The diagnosis of HAEC is often difficult to establish because clinical signs and symptoms are often non-specific in many patients. This difficulty often leads to over- and under-treatment of patients and it is also very difficult to compare the outcomes of different treatment strategies. An evidence-based rational approach to the diagnosis and management of HAEC is indispensable to reduce morbidity & mortality and to improve the quality of life of patients with HD.[3]

As it is very difficult to establish a definite diagnosis of HAEC, the incidence of HAEC varies widely, the reported incidence ranges from 6-60% prior to definitive pull-through surgery and from 25-37% after surgery.[3,4] Though all patients with Hirschsprung disease are at risk for HAEC, some factors appear to be associated with an increased risk. These include Down syndrome, long-segment aganglionosis, prior HAEC, and obstruction from any cause (retained aganglionosis, transition zone pull-through, dysmotility following pull-through, anastomotic stricture, twist in the pull-through or tight muscular cuff following the Soave procedure).[5]

1. Assistant Professor & Resident Surgeon, Division of Pediatric Surgery, Bangladesh Institute of Child Health, Dhaka Shishu (Children) Hospital.
2. Assistant professor, Division of Pediatric Surgery, Bangladesh Institute of Child Health, Dhaka Shishu (Children) Hospital.

Correspondence to: Dr. Md Ayub Ali, Assistant Professor, Bangladesh Institute of Child Health & Resident Surgeon, Dhaka Shishu (Children) Hospital. Cell: 01711143286, E-mail: ayub.ali.dhs@gmail.com
Received: 11 June 2018; **Accepted**: 12 July 2018

Etiology and Pathophysiology

The etiology of HAEC is yet to be known, despite numerous studies in this field. Several hypotheses have been proposed multifactorial occurrence based on experimental evidence. These include dysbiosis of the intestinal microorganism, impaired mucosal barrier function, altered innate immune responses, and bacterial translocation.[6-15]

Diagnosis

The classic presentations of HAEC include abdominal distention, fever, and diarrhea. However, the children present with a broad clinical spectrum, where other signs or symptoms are vomiting, per rectal bleeding, lethargy, loose stools, and obstipation. In fact, all these symptoms are non-specific and this likely contributes to the extensively variable incidence of HAEC reported in the literature. Mild cases, presenting with only fever, mild abdominal distension, and diarrhea are indistinguishable from viral gastroenteritis, which is very common in young children. On one side it is very difficult to make a definitive diagnosis, on the other side morbidity associated with a delayed or missed diagnosis is very high. Based on this fact, most pediatric surgeons deliberately over diagnose the cases of HAEC and presumptively treat suspected cases. Generally, it is wise to assume the child has HAEC and initiating treatment rather than delaying the diagnosis, which may progress later to more advanced disease.[5]

A group of 27 gastroenterologists and surgeons participated in a Delphi process, in an endeavor to reduce the difficulty in establishing the diagnosis of HAEC starting with 38 features (history, patient characteristics, physical exam signs, laboratory findings, radiology findings, pathology findings) and iteratively refining this list to 16 items to develop a HAEC score.[16] The resulting tool is clumsy and inconvenient for routine use but useful for standardizing outcome measures in research studies. This tool has not been validated for clinical application and has not been extensively adopted in the clinical setting. Another clinical grading system stratifies the degree of diarrhea, abdominal distention, and systemic presentations into mild, moderate and severe in order to assign an overall clinical grade. Following a staging system similar to that described by Bell for necrotizing enterocolitis (NEC),[17] and incorporating elements of published HAEC grading systems and clinical experience, the clinical suspicion, and severity of HAEC have been categorized into three grades based on history, physical examination, and imaging studies [Table I].

The goal of this approach is to produce a standard, clinically relevant, and convenient to use system that can be universally adopted to allow a consistent approach to the diagnosis and treatment of HAEC. In general, the presence of higher grade findings should prompt providers to err toward assigning the higher grade and initiating the corresponding treatment.[5]

Table I

Guideline for the diagnosis of HAEC

Grade	Description	Clinical History	Physical Examination	Radiologic findings
I	Possible HAEC	Anorexia, Diarrhea	Mild abdominal distention	Normal Mild ileus gas pattern
II	Definite HAEC	Past episode of HAEC Explosive diarrhea Fever lethargy	Fever Tachycardia Abdominal distention Abdominal tenderness Explosive gas/stool on DRE	Ileus gas pattern Air/fluid levels Dilated bowel loops Rectosigmoid cutoff
III	Severe HAEC	Obstipation Obtunded	Decreased peripheral perfusion Hypotension Altered mentation Marked abdominal distention Peritonitis	Pneumatosis Pneumo peritoneum

Table II

Guideline for the management of HAEC

Grade	Disposition	Diet	Antibiotics	Irrigations	Surgery
I	Outpatient	Oral hydration	Oral Metronidazole	Rectal irrigation	
II	Outpatient or Inpatient	Clear liquids or NPO IV hydration	Metronidazole (Oral/IV) Consider broad spectrum coverage	Rectal irrigations	
III	Inpatient, possible ICU	NPO IV hydration	Metronidazole Broad-spectrum coverage	Rectal irrigation	Proximal diversion for failure to improve with non-operative management Exploration for pneumo peritoneum

Management

As the cause of HAEC is mostly unknown, treatment usually goes on empiric and directed toward relieving acute symptoms as well as managing the contributing factors. The treatment guideline according to severity is summarized in Table II.[5]

In cases classified as Grade I (possible HAEC), outpatient management can typically be considered. Treatment in these cases should include oral metronidazole and oral hydration with an electrolyte-rich solution. Rectal irrigations should be considered in children who have abdominal distension or who have loaded rectum. Close monitoring is essential to detect symptoms progression to a higher grade of disease.

In more severe cases, Grade II (Definite HAEC), inpatient admission is frequently necessary. These children are managed usually with either clear liquids or nothing by mouth, intravenous fluids, and nasogastric decompression if there is significant abdominal distension. Rectal irrigations are very effective, which helps to resolve fecal stasis. Metronidazole (oral or parenteral) is used to treat anaerobes, including Clostridium difficile, which has been associated with HAEC. In addition, broad-spectrum intravenous antibiotic coverage using either the combination of ampicillin and gentamicin, or piperacillin/ tazobactam, or aztreonam (in the case of penicillin allergy) should be considered.

Children with Grade III (severe) HAEC, particularly with shock, may require admission to an intensive care unit. Bowel rest, intravenous fluid resuscitation, rectal irrigations, and broad-spectrum antibiotics (including metronidazole) are required. Proximal bowel diversion should be considered if symptoms don't improve. Rarely, pneumo peritoneum can occur, which would prompt immediate surgical intervention.[5]

Prevention

Some authors have advised the use of preventive measures in selected patient populations[2]. These measures include routine use of rectal irrigations in the post-operative period, long-term administration of oral metronidazole, and use of probiotic therapy. A recent prospective randomized trial found that 4 weeks of probiotic therapy decreased the incidence and severity of HAEC[18], but more studies are required.

Recurrent HAEC

Additional evaluation is required in children with recurrent HAEC to determine whether a cause can be identified. In these cases, an anatomic or pathologic cause of obstruction should be excluded. Anatomic problems include anastomotic stricture, a twisted or kinked anastomosis, mega rectum, or a tight Soave cuff. Causes of functional obstruction, like a transition zone pull-through or retained aganglionosis, should also be considered.[5]

Physical examination should include careful assessment for a stricture, presence, and function of the anal sphincters, size of the rectal pouch (if present), and presence of a palpable Soave cuff. It is

better to perform under anesthesia. A contrast enema using a water-soluble dye can identify any mechanical causes of obstruction. It should be avoided during acute HAEC episodes because of the risk of perforation. Further evaluation includes rectal biopsy to exclude aganglionosis or transition zone pull-through. Histopathology slides from the original surgery should be reviewed to exclude transition zone pull-through. If the evaluation reveals an anatomic etiology for obstructive symptoms and recurrent HAEC, surgical management should be considered to correct the defect. In absence of anatomic or pathologic causes, failure of relaxation of the internal anal sphincter may be the cause of stasis with obstructive symptoms and recurrent HAEC in some patients and require anorectal manometry to confirm the diagnosis.[5]

Clostridium botulinum toxin (Botox) injection into the intersphincteric groove has been shown to decrease hospital admissions in children with recurrent symptoms.[19]

References

1. Demehri FR, Halaweish IF, Coran AG, Teitelbaum DH. Hirschsprung-associated enterocolitis: pathogenesis, treatment and prevention. *Pediatric Surgery International* 2013; **29**(9): 873-81.
2. Frykman PK, Short SS. Hirschsprung-associated enterocolitis: prevention and therapy. *Semin Pediatr Surg* 2012; **21**(4): 328-35.
3. Haricharan RN, Seo J-M, Kelly DR. Older age at diagnosis of Hirschsprung disease decreases risk of postoperative enterocolitis, but resection of additional ganglionated bowel does not. *J Pediatr Surg* 2008; **43**: 1115-23.
4. El-Sawaf M, Siddiqui S, Mahmoud M. Probiotic prophylaxis after pullthrough for Hirschsprung disease to reduce incidence of enterocolitis: A prospective, randomized, double-blind, placebo-controlled, multicenter trial. *J Pediatr Surg* 2013; **48**:111-17.
5. Gosain A, Frykman PK, Cowles RA, Horton J, Levitt M, Rothstein DH et al. Guidelines for the Diagnosis and Management of Hirschsprung-Associated Enterocolitis. *Pediatr Surg Int.* 2017; **33**(5): 517-21.
6. Austin KM. The pathogenesis of Hirschsprung's disease-associated enterocolitis. *Seminars in Pediatric Surgery* 2012; **21**(4): 319-27.
7. Ward NL, Pieretti A, Dowd SE. Intestinal aganglionosis is associated with early and sustained disruption of the colonic microbiome. *Neurogastroenterology & Motility* 2012; **24**: 874-e 400.
8. Pierre JF, Barlow-Anacker AJ, Erickson CS. Intestinal dysbiosis and bacterial enteroinvasion in a murine model of Hirschsprung's disease. *J Pediatr Surg* 2014; **49**: 1242-51.
9. Yan Z, Poroyko V, Gu S, et al. Biochemical and Biophysical Research Communications. *Biochemical and Biophysical Research Communications* 2014; **445**: 269-74.
10. Frykman PK, Nordenskjöld A, Kawaguchi A. Characterization of Bacterial and Fungal Microbiome in Children with Hirschsprung Disease with and without a History of Enterocolitis: A Multicenter Study. *PLoS ONE* 2015;**10**:e0124172.
11. Thiagarajah JR, Yildiz H, Carlson T, et al. Altered Goblet Cell Differentiation and Surface Mucus Properties in Hirschsprung Disease. *PLoS ONE* 2014; **9**: e99944.
12. Yildiz HM, Carlson TL, Goldstein AM, Carrier RL. Mucus Barriers to Microparticles and Microbes are Altered in Hirschsprung's Disease. *Macromol Biosci* 2015; **15**: 712-18.
13. Gosain A, Barlow-Anacker AJ, Erickson CS. Impaired Cellular Immunity in the Murine Neural Crest Conditional Deletion of Endothelin Receptor-B Model of Hirschsprung's Disease. *PLoS ONE* 2015; **10**: e0128822.
16. Frykman PK, Cheng Z, Wang X, Dhall D. Enterocolitis causes profound lymphoid depletion in endothelin receptor B- and endothelin 3-null mouse models of Hirschsprung-associated enterocolitis. *Eur J Immunol* 2015; **45**: 807-17.
15. Cheng Z, Dhall D, Zhao L, et al. Murine model of Hirschsprung-associated enterocolitis.I: Phenotypic characterization with development of a histopathologic grading system. *J Pediatr Surg* 2010; **45**: 475-82.
16. Pastor AC, Osman F, Teitelbaum DH, et al. Development of a standardized definition for Hirschsprung's-associated enterocolitis:a Delphi analysis. *J Pediatr Surg* 2009; **44**: 251-56.
17. Bell MJ, Ternberg JL, Feigin RD, et al. Neonatal necrotizing enterocolitis. Therapeutic decisions based upon clinical staging. *Ann Surg* 1978; **187**: 1-7.
18. Wang X, Li Z, Xu Z, et al. Probiotics prevent Hirschsprung's disease associated enterocolitis: a prospective multicenter randomized controlled trial. *Int J Colorectal Dis* 2015; **30**: 105-10.
19. Patrus B,Nasr A, Langer JC,Gerstle JT. Intrasphincteric botulinum toxin decreases the rate of hospitalization for postoperative obstructive symptoms in children with Hirschsprung disease. *J Pediatr Surg* 2011;**46**:184-87.

DS (Child) H J 2018; 34(1) : 123-126

CASE REPORT

Agenesis of Left Lung with Ostium Secundum ASD: A Rare Report

Nabila Akand[1], Probir Kumar Sarker[2], Md. Kamruzzaman Kamrul[3], Sakib Rana[4], Khondokar Ashiq-Ur-Zaman[5]

Introduction

Pulmonary agenesis is a very rare congenital abnormality that represents the unsuccessful growth of the primitive lung bud.[1] Agenesis means complete absence of bronchus, lung parenchyma and associated vessels.[2] Bilateral pulmonary agenesis is incompatible with life, manifesting as severe respiratory distress and failure. Pulmonary agenesis is thought to be an autosomal recessive trait, with an estimated incidence of 1 in 10,000-15,000 births.[3]

The pathogenesis of such anomalies is not accurately known with several proposed theories. Almost half of pulmonary agenesis cases accompany congenital defects, which may involve multiple organ systems.[4] It is often associated with anomalies of the cardiovascular, genitourinary systems. An isolated left lung agenesis is rare and more commonly involves the right upper and middle lobes.[5] We describe here an infant who was shown to have agenesis of the left lung with a small ostium secundum ASD.

Case Report

Sami a 1 month old male young infant, 3rd issue of his parents got admitted to this hospital at 24th day of age with the complaints of cough and respiratory distress since 12th day of his life. The respiratory distress was not associated with feeding, recurrent vomiting or blueish discoloration. With these complaints initially he was treated in a tertiary level hospital and was referred for further evaluation and better management. The baby was delivered at term at hospital with average birth weight. On examination, the baby was ill looking, dyspnoeic with inter-costal and sub-costal chest recession, R/R-65/min, temp-98° H/R-150/min. There was no cyanosis, oedema or dehydration. His weight -2.3Kg, length- 50cm and OFC- 34cm. His chest was depressed on the left side and chest movement was also decreased on the ipsilateral side. The mediastinum was found to be shifted on the left side evidenced by shifting of trachea to the left and apex beat located on the left 4th intercostal space just lateral to the mid-clavicular line. Percussion note was dull on the left side but was resonant on the right side and breath sound was absent on the left hemi-thorax and was vesicular with diffuse crepitations on the right side. There was no organomegally and other systemic examinations were normal. So provisionally the baby was diagnosed as a case of left sided collapse-consolidation.

CBC values were normal except for thrombocytopenia. The serial chest radiographs anterior-posterior view Figure 1 revealed volume loss of left lung field with the features of collapse evidenced by ribs crowding on the left side and elevation of the left hemidiaphragm. Secondary changes were seen in the form of hyperinflated right lung, herniation across the midline, and shift of mediastinum to the left.

1. Registrar, Department of Pediatric Pulmonology, Dhaka Shishu (Children) Hospital.
2. Associate Professor & Head, Department of Pediatric Pulmonology, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital.
3. Assistant Professor, Department of Pediatric Pulmonology, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital.
4. Honorary Resident Medical Officer, Department of Pediatric Pulmonology, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital.
5. Resident Medical Officer, Department of Pediatric Pulmonology, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital.

Correspondence to: Dr. Nabila Akand, Registrar, Dhaka Shishu (Children) Hospital. Cell: 01670002801, E-mail: nabilahanahasib@gmail.com

Received: 17 April 2018; **Accepted**: 22, May 2018

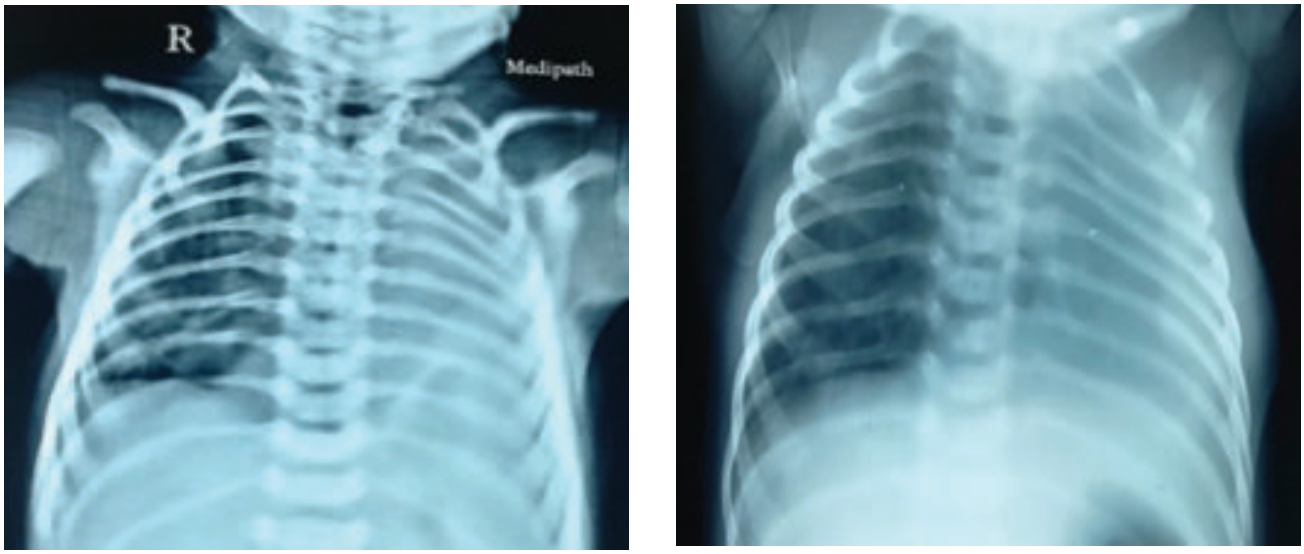

Fig 1 Serial Chest X-rays

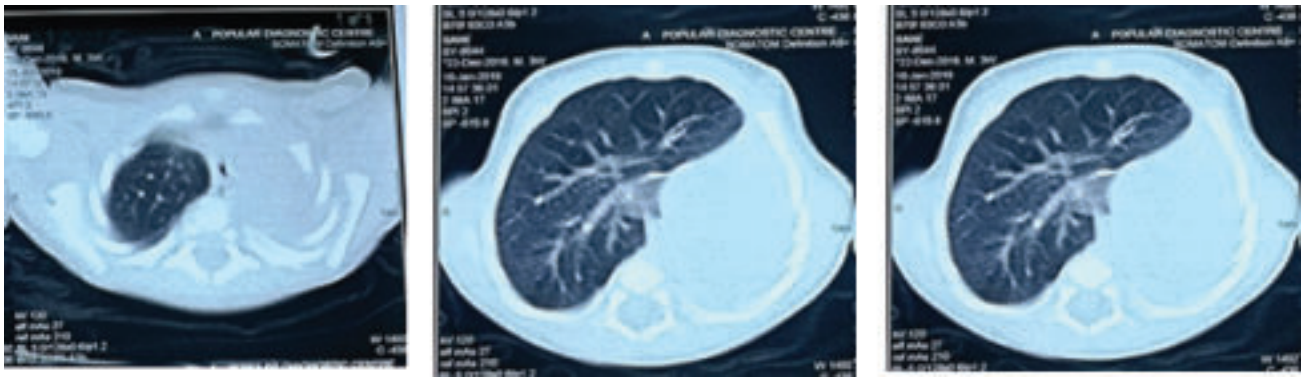

Fig 2 CT scan of the chest

After that chest CT scan was performed. CT scan of chest Figure 2 revealed that the left lung was not visualized. Compensatory hyperinflation on the right side with herniation of the right lung across the midline was also seen.

On colour droppler echocardiography, small ostium secundum ASD was found. Main pulmonary artery continuous as right pulmonary artery and left pulmonary artery was non-visualized. No other associated congenital anomaly was found on ultrasonography of abdomen. Finally, the patient was diagnosed as a case of agenesis of left lung with small ostium secundum ASD.

He was initially treated with parenteral antibiotics with supportive care for 7 days. There after surgical consultation was also taken. The chest surgeon has opted for continuing conservative treatment. Counselling was done to his parents about the congenital pulmonary malformation of his airways and cardiovascular anomalies and thereafter was kept under follow up.

Discussion

Pulmonary agenesis is one the rarest congenital disorders of the lung. The other forms of congenital disorders of the lung includes aplasia, hypoplasia, congenital cystic malformation, pulmonary sequestration, bronchogenic cysts, lung hernia, congenital pulmonary lymphangiectasia, congenital lobar emphysema and pulmonary cyst, arteriovenous malformation. Among them some authors have classified 'underdevelopment of lung' into three catagories:

1. Agenesis shows complete absence of bronchus, lung parenchyma and associated vessels.

2. Aplasia has a rudimentary bronchus with absent lung parenchyma.
3. Hypoplasia has hypoplastic bronchus and lung parenchyma.[6]

It differs from hypoplasia in that agenesis means the complete absence of a lung. Agenesis differs from aplasia by the absence of a bronchial stump or carina that is seen in aplasia.[3] Pulmonary agenesis has been classified morphologically based on the extent to which broncho-pulmonary tissue is absent. Spencer modified earlier classification and divided pulmonary agenesis into three catagories: (1) bilateral complete agenesis; (2) unilateral agenesis; and (3) lobar agenesis or lesser forms of congenital anomaly.[7]

The etiopathogenesis is unclear; however, genetic, teratogenic agents (allopurinol), and Vitamin A deficiency during pregnancy have been hypothesized as its causes.[8,9]

The abnormality goes undetected or is incidentally detected in the majority of cases. Symptomatic patients present in early childhood with symptoms of recurrent respiratory tract infection while others remain asymptomatic. Underdevelopment of lung is usually associated with other congenital]abnormalities of cardiovascular, musculoskeletal, and genitourinary system, more commonly toward the ipsilateral side. Mardini-Nyhan association is seen in consanguineous marriages, comprising congenital heart disease, thumb abnormality, and complete/partial lung agenesis.[10] Our case had associated small ostium secundum with mild right pulmonary artery stenosis.

Genetic defects of the pulmonary system and embryonic disorders of the bronchi contribute to anomaly, but the pathogenesis remains controversial. Less than 5% of the general population develop bronchial anomalies, and the malformation of the right upper bronchus is the most common abnormality. Pulmonary artery hypoplasia is commonly accompanied with such an anomaly of the lung.[11] An investigation of the associated vascular abnormalities is therefore necessary.

Isolated lobar agenesis is usually asymptomatic and may remain undiagnosed throughout childhood until a referral for an abnormal chest radiograph.[12] In left lung agenesis, the presentations of a chest radiograph include a decreased left lung volume, shifting of the mediastinum on the left side, and an elevation of the left hemidiaphragm. The differential diagnosis of our patient's chest radiograph includes left lung atelectasis, pulmonary hypoplasia and left sub-pulmonic effusion. A chest CT is considered to be the most conclusive examination to diagnose congenital abnormalities of the lung and the associated vascular anomalies when a chest radiograph is not diagnostic.[13] Three-dimensional reconstruction can be particularly helpful in delineating abnormalities of the bronchi and associated arterial and venous structures. Angiography is reserved for patients requiring embolization or revascularization surgery. Treatment is necessary for recurrent chest infections. The prognosis is good if there is no other malformation and if the diagnosis has been incidental.[2]

Conclusion

An infant who has been consecutively demonstrating an abnormal chest radiograph with or without respiratory symptoms, we should always consider a congenital anomaly of the lung. CT scan of chest may be useful in determining the extent of the lesion and to define associated abnormalities.

References

1. Mardini MK. Nyhan WL. Agenesis of the lung: report of four patients with unusual anomalies. *Chest*. 1985; **87**: 522-27.
2. Gupta K, Taneja D, Aggarwal M, Gupta R. Left upper lobe agenesis of lung: A rare case report. *Lung India* 2017; **34**(3): 302-04.
3. Joshua A. Blatter and Jonathan D. Finder editor. Nelson Textbook of Paediatrics: Congenital Disorders of the Lung. 20th Edition. Vol 2. Philadelphia: Elsevier; 2016. p.2057.
4. Chiu-Ping K, Yen-Ta L, Rong-Luh L. Agenesis of right upper lobe. *Respiratory Case Reports* 2015; **3**(2): 51-53.
5. Clements BS. Congenital malformations of the lungs and airways. In: Landau LI, Taussing LM, editors. *Pediatric Respiratory Medicine*. St. Louis, MO, USA:Mosby; 1999. pp.1106-36.
6. Berrocal T, Madrid C, Novo C, Gutiërrez J, Arjonilla A, Gymez-Leyn N. Congenital anomalies of the tracheobronchial tree, lung, and mediastinum: Embryology, radiology and pathology. *Radiographics* 2004; **24**: e17.

7. Schwartz MZ , Ramchandran P. Congenital malformation of the lung and mediastinum- a quarter century of experience from a single institution. *J Pediatr Surg* 1997; **32**: 44-47.

8. Kozenko M, Grynspan D, Oluyomi-Obi T, Sitar D, Elliott AM, Chodirker BN. Potential teratogenic effects of allopurinol: A case report. *Am J Med Genet A* 2011; **155**A: 2247-52.

9. Kisku KH, Panigrahi MK, Sudhakar R, Nagarajan A, Ravikumar R, Daniel JR. Agenesis of lung – A report of two cases. *Lung India* 2008; **25**: 28-30.

10. Hastings R, Harding D, Donaldson A, Liebling R, Hayes A, Kraus A, et al. Mardini-Nyhan association (lung agenesis, congenital heart, and thumb anomalies): Three new cases and possible recurrence in a sib-is there a distinct recessive syndrome? *Am J Med Genet A* 2009; **149**A: 2838-42.

11. Stovin PGI. Early lung development. *Thorax*. 1985; 40: 401-05.

12. Zack MS, Eber E. Adult outcome of congenital lower respiratory tract malformations. *Thorax* 2001; **56**: 65-72.

13. Kuo CP, Lu YT, Lin RL. Agenesis of right upper lobe of lung. *Respirol Case Rep* 2015; **3**: 51-53.

DS (Child) H J 2018; 34(1) : 127-129

CASE AREPORT

Rare Presentation of Cystic Fibrosis Arthropathy - A Case Report

Khan Lamia Nahid[1], Md. Rukunuzzaman[2], Rana Kumar Biswas[3], Fahmida Begum[1], ASM Bazlul Karim[5]

Introduction

Cystic fibrosis (CF) is an autosomal recessive disease which is uncommon condition among South East Asian population. It is even less reported in Bangladesh because of lack of diagnostic facilities. Cystic fibrosis is a life-threatening genetic disease for Caucasian population. In the United States, cystic fibrosis (CF) occurs in approximately 1:3000 Caucasians, 1:9200 Hispanics, 1:10,900 Native Americans, 1:15,000 African Americans, and 1:30,000 Asian Americans.[1] The median predicted survival for CF patients in the United States was 39.3 years (95% CI, 37.3-41.4) according to the Cystic Fibrosis Foundation 2014 Registry Report.[2] CF is increasingly recognized among non-white population, not only in regions familiar with CF but also in South and East Asia. But prevalence in this region is not studied extensively. It is diagnosed mainly by typical clinical features and positive sweat chloride test.[3] Detection of Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene is not possible in our country. It is mainly a disease of pulmonary and gastrointestinal systems. Other systems are rarely involved. There are a number of well known rheumatic conditions that may affect patients with CF. these are Cyctic fibrosis arthropathy, hypertrophic osteoarthropathy, osteoporosis, vasculitis (cutaneous, systemic), Juvenile rheumatoid arthritis, sarcoidosis, psoriatic arthritis, fluoroquinolone-induced arthropathy, bowel disease related arthritis etc. Cystic fibrosis arthropathy is an episodic, acute arthropathy which affects 2-8% of adults with cystic fibrosis. It usually affects large joints, mainly non-erosive in nature and responds rapidly by NSAIDS. It rarely occurs below 10 years of age.[4] The prevalence of Cystic fibrosis arthropathy in children is not well known. It is of unknown etiology, although it is thought to relate to circulating immune complexes.

Case Report

A six and half years old boy of consanguineous parents presented in the department of Paediatric Gastroenterology & Nutrition of BSMMU on December, 2017 with cough for fifteen days which was productive in nature, associated with fever, chest pain and respiratory distress. His mother also complaints of pain and swelling of multiple large and small joints of hands and legs for last three weeks which was additive in nature, not associated with morning stiffness, non-migratory in nature and hampered daily activities. On query, his mother mentioned that her child has been suffering from recurrent respiratory tract infection and passage of bulky oily stool since birth. He had no history of rash,

1. Assistant Professor, Department of Pediatric Gastroenterology and Nutrition, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.
2. Associate professor, Department of Pediatric Gastroenterology and Nutrition, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.
3. MD Resident Phase B, Department of Pediatric Gastroenterology and Nutrition, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.
4. Professor, Department of Pediatric Gastroenterology and Nutrition, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.

Correspondence to: Dr Khan Lamia Nahid, Assistant Professor, Department of Pediatric Gastroenterology % Nutrition, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh. Email: lamianahid@yahoo.com
Received: 14 May 2018; **Accepted**: 17 June 2018

bleeding manifestations, oral ulceration or contact with tuberculosis patient. He had repeated hospital admission for respiratory tract infection. At the age of one year, he was diagnosed as a case of cystic fibrosis on the basis of typical clinical features and sweat chloride test. He had no positive family history. Mutation analysis of CFTR gene was not possible in our country. He received different types of antibiotics and nebulization during each hospital course. On general examination, the boy was fretful, mildly pale and generalized clubbing was present. His vitals were normal, but was moderately under-weight and wasted. His BMI for age is less than 5th centile. On respiratory system examination, respirato ry rate was high (52 breath/min), chest indrawing was present and breath sound was vescicular in nature. Bilateral crepitations were present in both lung fields. Gastrointestinal system examination revealed hepatomegaly. Locomotor system revealed swelling of both knee, ankle and wrist joints with increased temperature of affected joints and tenderness grade 4 over 5. There was no joint deformity. Patellar tap and fluctuation test were positive. Antalgic gait was present.

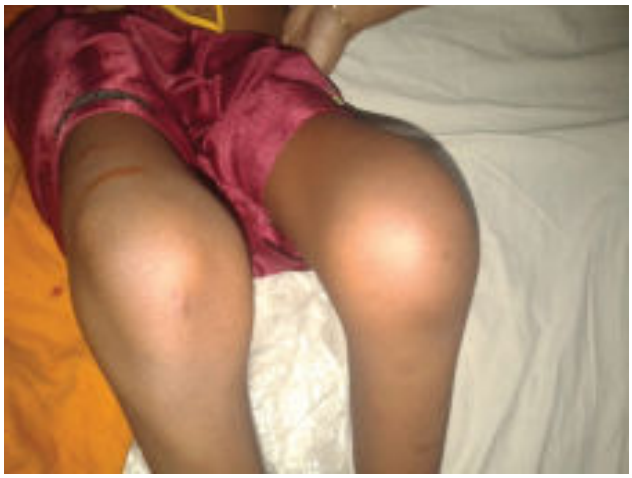

Fig 1 *Swelling of both knee joints*

On lab investigations, CBC showed mild anaemia, thrombocytosis and mild leukocytosis. ESR was moderately raised. Liver function test showed normal findings. Stool for fecal fat was 60-100 droplet/HPF and sweat chloride was 110 and 90 mmol/L for two separate occasion. X-ray of knee joint showed enlarged joint space. Antinuclear antibody and serum rheumatoid factor were negative. He was treated with non steroidal anti inflammatory drugs (naproxen, 10 mg/kg/day) for 10 days. Pain and swelling was subsided gradually. During this period, no immunsuppressive agents were required. We followed up the patient subsequently on three months interval for one year which showed no recurrence.

Discussion

Cystic Fibrosis Arthropathy (CFA) is a unique manifestation of Cystic fibrosis. It was first reported in 1979 by Ansell and Newman,[5] that usually starts after ten years of age, occurring in about five to ten percent of patients.[6] It is a specific condition, which may be immune-complex mediated and related to chronic pulmonary infection and inflammation. Typically, the children have episodic arthritis that usually involves large joints such as knees, ankles and wrists. It is often accompanied by low-grade fever and there may be erythematous rashes or purpura.[6,7] Joint x-rays are usually normal but may have effusion. Although in most cases there is no erosion to the affected joints, in a few cases the condition may progress to erosive arthritis with bone destruction[7]. Episodes tend to decrease spontaneously after 7 days and respond well to non-steroidal anti-inflammatory drugs. However, episode of joint inflammation may last several weeks in some cases. The arthritis follows a remitting and relapsing course. The patient is completely normal in between attacks. The majority of reported patients are negative for both serum rheumatoid factor and antinuclear antibody.[7-9] CFA is not early feature for CF, usually occur later than the pulmonary or gastrointestinal manifestations of CF. While CFA has been reported in a number of patients under 6 years of age[4], the average age of CFA patients at diagnosis in the 3 surveys was 17 years.[10] Three groups have attempted to survey their patient populations, with varying success. In 1986, Rush et al[11] described 12 cases of CFA from among 533 CF patients at their hospital (2.3%). Bourke and colleagues prospectively surveyed all 59 of their CF patients ages >14 years for the presence of arthritis symptoms and then clinically evaluated those with positive findings. They found 8.5% prevalence of CFA.[12]

A second form of bone and joint disease is called hypertrophic pulmonary osteoarthropathy (HPOA). It is usually found in young adult patients and has a slow onset. Pain, which is generally mild at the beginning, may increase gradually. The clinical picture may vary from a minimally swollen joint to

tender, warm and swollen joints resembling those seen in rheumatoid arthritis. Symptoms are often aggravated during cold weather. It manifests as swelling of the ends of the long bones. In most patients the joint symptoms are symmetrical such as both wrists, both knees or both ankles. X-rays show the periosteum may be elevated from the bone surface and eventually new bone formation occurs. These x-ray changes appear later in the disease course. The changes can slowly progress and can result in permanent bony destruction. Both the above forms of arthritis can be associated with acute respiratory exacerbations. This may reflect the upgraded immune response that accompanies acute infections, the arthritis being another manifestation of a hyperactive immune system.

In our case, the boy suffered from pain and swelling of multiple joints which lasted for several days. He was diagnosed having Cystic fibrosis (CF) earlier in life. His main problems are respiratory and gastrointestinal, which started early. His joint symptoms developed later. His clinical feature were consistent with Cystic fibrosis Arthropathy (CFA). Though age of onset is early in this case.

Usually CFA tends to occur after ten years of life. Both large and small joints were involved in this case without any deformity. The patient was negative for both serum rheumatoid factor and antinuclear antibody which matched with the previously described cases. The joint manifestations flare up with acute respiratory exacerbations. It responded well with nonsteroidal anti-inflammatory drugs. Immuno-suppressive agents were not used here. Treatment of rheumatic conditions in CF is not well defined. Immunosuppressive agents are used less often in case of CFA.

Conclusion

Cystic fibrosis arthropathy is a rare condition for paediatric population. Though it is common among all rheumatic manifestations of CF. Cystic fibrosis patients may have various nonspecific musculo-skeletal problems such as chest pain and back pain due to chronic coughing. Seropositive Juvenile rheumatoid arthritis may have association with CF. CFA responds well with NSAIDs , if not responds well with NSAIDs, then search for other rheumatic conditions are necessary.

References:

1. Hamosh A, FitzSimmons SC, Macek M Jr, Knowles MR, Rosenstein BJ, Cutting GR. Comparison of the clinical manifestations of cystic fibrosis in black and white patients. *J Pediatr* 1998; **132**: 255.
2. Cystic Fibrosis Foundation Patient Registry: Annual Data Report to the Center Directors, 2014. https://www.cff.org/2014_CFF_Annual_Data_Report_to_the_Center_Directors.pdf/ (Accessed on December 11, 2015).
3. Farrell PM, White TB, Ren CL, Hempstead SE, Accurso F, Derichs N, et al . Diagnosis of Cystic Fibrosis: Consensus Guidelines from the Cystic Fibrosis Foundation. *J Pediatr* 2017; 181S: S4-S15.e1.
4. Dixey J, Redington AN, Butler RC, Smith MJ, Batchelor JR, Woodrow DF, et al. The arthropathy of cystic fibrosis. *Ann Rheum Dis* 1988; **47**: 218-23.
5. Newman AJ, Ansell BM. Episodic arthritis in children with cystic fibrosis. *J Pediatr* 1979; **94**: 594-6.
6. Lawrence JM, Moore TL, Madson KL, et al. Arthropathies of cystic fibrosis: case reports and a review of the literature. *J Rheumatol* 1993; **20** (suppl 38): 12-15.
7. Wulffraat NM, de Graeff-Meeder ER, Rijkers GT, van der Laag H, Kuis W. Prevalence of circulating immune complexes in patients with cystic fibrosis and arthritis. *J Pediatr* 1994; **125:** 374-8.
8. Jones SM, McKay KM, McHugh NJ. Bilateral hip replacements in a man with cystic fibrosis and episodic arthritis. *Br J Rheumatol* 1997; **36**: 298-9.
9. Taccetti G, Campana S, Marianelli L, Turchini S. Cystic fibrosis and episodic arthritis [letter]. *J Pediatr* 1995; **126**: 848-49.
10. Pertuiset E, Menkes CJ, Lenoir G, Jehanne M, Douchain F, Guillot M. Cystic fibrosis arthritis: a report of five cases. *Br J Rheumatol* 1992; **31:** 535-38.
11. Rush PJ, Shore A, Coblentz C, Wilmot D, Corey M, Levison H. The musculoskeletal manifestations of cystic fibrosis. *Semin Arthritis Rheum* 1986; **15**: 213-25.
12. Bourke S, Rooney M, Fitzgerald M, Bresnihan B. Episodic arthropathy in adult cystic fibrosis. *QJM* 1987; **64**: 65-59.

DS (Child) H J 2018; 34(1) : 130-132

CASE REPORT

Gaucher Disease with Pregnancy - A Successful Outcome

Umme Parveen[1], Shahin R Chowdhury[2], Farid Ahmed[3]

Introduction

Gaucher disease is an autosomol recessive lysosomal storage disease resulting from deficiency of enzyme glucocerebrosidase which is required for lysosomal degradation of glycolipids. The disease occurs in three clinical forms. Type 1 (adult) is by far the most common type; it is characterized by accumulation of glucocerebroside in the spleen, liver, and bone marrow and by sparing of the central nervous system. About 50-60% of such patients are diagnosed before 20 years of age. Type 2 (infantile) and type 3 (juvenile) are the two rare neuropathic forms. Gaucher disease type 2 is a rare, severe CNS disease that leads to death by 2 years of age. Type 3 gaucher disease is intermediate form has highly variable manifestations in the CNS & visera. Type I disease affects 1 in 50.000-100.000 people worldwide and 1 in 400-600 people among Ashkenazi Jewish population.[1]

Disease variants are classified based on the absence or presence and progression of neuronopathic involvement. Gaucher disease is a rare condition affecting the reticulo-endothelial system. Enzyme deficiency results in accumulation of Glucocerebroside within the reticulo-endothelial system. This is characterized by and enlargement of spleen some time reaching to enormous dimension, less marked hepatomegaly, moderate anaemia, leucopenia and thrombocytopenia. There is a tendency to bleeding, skeletal changes, a brownish pigmentation of skin and a peculiar yellowish thickening of the conjunctiva. The disease has a familial tendency and may appear in infancy and childhood but diagnosis is usually at adulthood. Females are more often affected than males and the disease has a predilection for Jewish race.[2]

Few data on the prevalence of Gaucher disease during reproductive age and pregnancy can be found in the literature. Prevalent data have been reported in Netherlands (2 per 100,000 live births) in Australia (1 per 57,000 live births) portugal (3 per 100,000 live births).[3]

Case report

A 22 years old primigravida patient hailing from Dhaka presented at 33 weeks of gestation with moderate anaemia, weakness and pregnancy induced hypertension. She was a known case of Gaucher disease type 1. She was diagnosed one and half year back. She remained asymptomatic until the age of 20 when she developed generalized fatigue and bleeding tendency. Her CBC showed pancytopania and grossly enlarged liver and spleen. At that time her CBC report was Hb% 8.7gm/dl, RBC 3.45x10^{Ù}12/L, platelet count 80x10^{Ù}9/L, WBC 2.50x10^{Ù}9/L. Blood film report showed anisopoikilocytosis and anisochromia. WBC and platelet count was reduced. Hb Electrophoresis showed normal study.

Her duplex study of hepatobilliary system showed Echogenic hepatic paranchyma, Splenomegaly and borderline dilated splenic vein. Spleen was markedly enlarged (27cm x 13.5cm), CT scan of whole Abdomen showed gross hepatosplenomegaly (liver size 20.8 cm, spleen size 26 cm). Then she was advised for Bone marrow study. Bone marrow report showed hypoplastic marrow. She was advised for Bone

1. Assistant Professor, Department of Obstetric and Gynaecology, Holy Family Red Crescent Medical College, Dhaka.
2. Professor, Department of Obstetric and Gynaecology, Holy Family Red Crescent Medical College, Dhaka.
3. Professor and Head of General Pediatrics, Bangladesh Institute of Child Health and Dhaka Shishu Hospital.
Address of correspondence: Dr. Umme Parveen, Assistant Professor, Department of Obstetric and Gynaecology, Holy Family Red Crescent Medical College, Dhaka. Cell: 01713035515, E-mail: ummeparveen@live.com
Received: 17 July 2018; **Accepted**: 21 August 2018

marrow trephine biopsy. Trephine biopsy revealed Gaucher disease. Section showed spongy bone. Marrow showed large collection of enlarged histiocytes. These cells had abundant fibrillary mildly PAS positive cytoplasm. Erytroid myeloid precursors appeared normal. Mild fibrosis was present. X-Ray of pelvis with both femurs showed normal study. Endoscopy of upper GI tract revealed gastritis. Finally she was advised for splenectomy. Within 1 year of diagnosis she got married and was pregnant soon after. She was on infrequent antenatal checkup.

At 16 weeks pregnancy she did a Hb level which was 8.8 g/dl. USG showed 16 weeks live pregnancy with massive spleenomegaly. At 33 weeks pregnancy CBC was done which revealed pancytopania. Her urine for Albumin was trace and urine sugar was nil. USG revealed 33 weeks live pregnancy with cephalic presentation with gross splenomegaly. Enormous spleen did not interfere with the normal increase in uterine size. Her BP was 125/85 mm Hg. She was advised for admission at 36 weeks as her BP was 160/100 mm Hg. Hb 8.9 gm/dl, platelet 48x$10^{Ù}$9/L, WBC 2.7X$10^{Ù}$9/L, RBC 3.6X$10^{Ù}$12/L, uric acid 5.9 mg/dl, Alkaline phosphatase 186 U/L, urinary albumin trace. RBS, LDH, S. creatinine, SGPT was within normal range, Antihypertensive drug was started and one unit of blood was transfused. USG revealed cephalic presentation, gross hepato splenomegaly, features of portal hypertension and Bilateral hydronephrosis. Single episode of convulsion occurred and Inj. $MgSO_4$ was given. Emergency LUCS done after collaborative consultation with internal medicine, ICU and Anaesthesia department. Very carefully LUCS was under spinal anesthesia was done as because lower limit of spleen reached upto the uterine incision. Gently foetus was delivered to avoid splenic rupture. A 2.74 kg male child with good APGAR SCORE was delivered. Per operatively 1 bag blood and 1 bag of Apheresis platelet was transfused. After LUCS Patient send to ICU. At ICU patient received injection Filasin (Human Granulocyte colony stimulating factor), Inj. Amikacin, Inj Metronidazole, Inj Brodactum, Inj. Omeprazol, Inj. Emistate, Inj. 20% Albutin.

On 2nd POD she received 1 unit of whole blood and patient transferred to cabin. On 6th POD CBC showed Hb% 10.8 gm/dl RBC 3.72 X $10^{Ù}$12/L, platelet 95 X $10^{Ù}$9/L, WBC 3.7 x$10^{Ù}$9/L. On 9th POD she was normotensive and baby was well and discharged with advice. There was minimal amount of P/V bleeding during post partum period. The lactation was normal. The Gaucher Disease did not adversely affect the course of pregnancy and there was no change in size

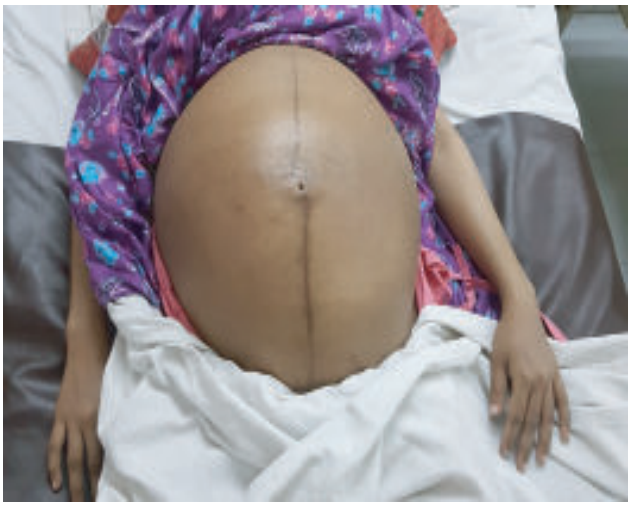

Fig 1 35 weeks pregnancy with hepatosplenomegaly

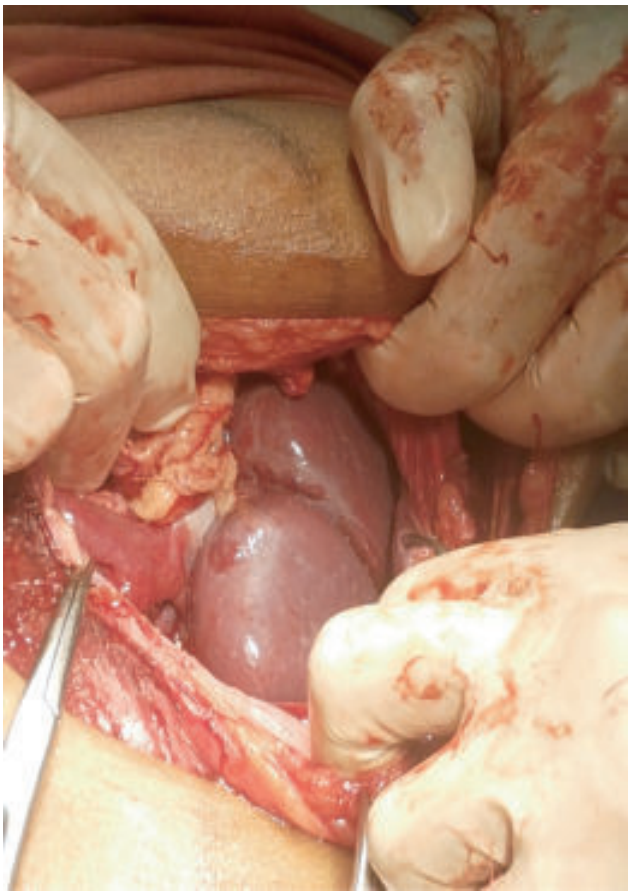

Fig 2 Lower border of spleen reaches upto the uterine incision.

of liver and spleen after pregnancy. Patient was advised to maintain contact with medicine specialist. She was also advised to seek surgical opinion regarding necessity for splenectomy.

Fig 3 Baby with her mother before discharge

Discussion

Gaucher disease is not very frequently seen in pregnancy. When pregnancy complicates the condition a decision must be made about termination of pregnancy. There is anxiety of possible rupture of spleen during stress of labour and mechanical interference with the natural uterine enlargement. In addition large liver may be damaged by the metabolic changes and demands of pregnancy.

Bromberg et al[4] in their study of nine pregnant woman with gaucher disease found in every case there was comfortable pregnancy followed by successful normal full term delivery. In one case there was opportunity to observe the patient over a period of 7 years. No detoriation in her condition observed.

Addleman etal[5] in one study considered abortion for a patient because she had 2 children. But patient had strong desire to continue pregnancy and outcome was good.

In our study pregnancy was not much affected though there was huge splenomegaly. Normal uterine enlargement during pregnancy was unaffected. Though there was pancytopenia post operative period was uneventful. An average size healthy baby was delivered. There was no evidence of haemorrhage from spleen or liver. Puerperum was normal and patient was discharged on 9th day of normal puerperum. The general impression received during this relatively short period of observation was that pregnancy have no adverse effect upon the disease.

In one case report Giannubilo et al[6] showed recombinant human enzyme (imiglucerase) treatment throughout the pregnancy. On that study no congenital anomalies or adverse neonatal outcome were registered. In my case no drug was used during pregnancy and there was no congenital anomalies.

Conclusion

The compatibility of Gaucher disease and pregnancy has been established. This case gives impression about relative benign nature of the disease in adult female. The conclusion drawn that conception in women with gaucher disease should not be discouraged. Normal pregnancy can be carried to term in presence of hepatosplenomagaly. In the absence of complications abortion is not justified.

References

1. Cox TM, Schofield JP. Gaucher's disease: clinical features and natural history. *Baillieres Clin Haematol.* 1997; **10**(4): 657-89.
2. Elstein Y, Eisenberg V, Granovsky-Grisaru S, Rabinowitz R, Samueloff A, Zimran A, et al. Pregnancies in Gaucher disease: a 5-year study. *Am J Obstet Gynecol.* 2004; **190**(2): 435-41.
3. Pinto R, Caseiro C, Lemos M, Lopes L, Fontes A, Ribeiro H, et al. Prevalence of lysosomal storage diseases in Portugal. *Eur J Hum Genet.* 2004; **12**(2): 87-92.
4. Bromberg YM, Toaff R, Diengott D. pregnancy with Gaucher disease. *British Medical Journal.* 1953; **2**: 761-63.
5. Addleman W, Gold S. Case Report: Gaucher disease and pregnancy. *Med Ass J.* 1963; **89**: 821-23.
6. Giannubilo SR, Pasculli A, Tidu E, Ciavattini A. Replacement therapy for Gaucher Disease during pregnancy: A case report. *J Reprod Infertil.* 2015; **16**(1): 53-57.

DS (Child) H J 2018; 34(2) : 133-134

ABSTRACTS FROM CURRENT LITERATURE

The Correlation Between Inferior Vena Cava Diameter Measured by Ultrasonography and Central Venous Pressure

Hans Vaish, Virendra Kumar, Rama Anand, Viswas Chhapola, Sandeep Kumar Kanwal

The Indian Journal of Pediatrics October 2017, Volume 84, Issue 10, pp 757-762

Objective : To find a correlation between inferior vena cava (IVC) diameters, IVC compressibility index (CI) and central venous pressure (CVP).

Methods : Prospective observational study was done at pediatric intensive care unit (PICU) of Kalawati Saran Children's Hospital (KSCH). Fifty children aged 5-18 y, presenting with shock were enrolled for the study. IVC diameters, CI and relevant clinical data were noted at enrollment, 30 min, 1 h, 6 h, and 12 h. Central line was placed at the time of admission.

Results : Of 50 children enrolled, 28 were boys, with a mean age of 11 y. More than 80% of cases were diagnosed as septic shock. Mean maximum and minimum IVC diameter of 8.3 ± 2 mm and 3.7 ± 1.7 mm, respectively CI 58.2 ± 7% and CVP of 5.4 ± 1.5 cm of H2O was observed at admission. CVP and IVC diameters showed a serial improvement with treatment; CI showed a serial decrease with treatment. Heart rate (HR) and systolic blood pressure (SBP) also showed a serial improvement at 12 h ($p < 0.05$). CVP showed a positive correlation with IVC diameter (r +0.312; $p < 0.05$), and a negative correlation with CI (r -0.343; $p < 0.05$).

Conclusions : Effective fluid resuscitation improves IVC diameters with a decrease in CI. IVC diameter has a positive correlation to CVP and CI has a negative correlation to CVP.

Clinical Presentation and Cardiovascular Outcome in Complete versus Incomplete Kawasaki Disease

G Shivalingam, GP Prashanth, Kallesh Hebbal, Rodney Aguiar

Indian Pediatrics. October 15, 2017; 54: 844-47.

Objective: To compare the demographic, clinical, and laboratory features of incomplete and complete presentations of Kawasaki disease.

Methods: A retrospective review of the electronic case records between January 2000 and December 2015 in a tertiary care referral center of Sohar, Oman.

Results: 31 out of 64 children (48.4%) had incomplete presentation. Children with incomplete presentation had higher incidence of skin rash, lymphadenopathy and conjunctivitis. They took a longer time to show clinical response to intravenous immunoglobulin [mean (SD) 52.6 (17.4) h *vs* 40.1 (16.4) h, *P*=0.005], and had prolonged hospitalization [mean (SD) 6.2 (2.5) d *vs* 4.6 (1.7) d, *P*=0.009].

Conclusion: Children with incomplete presentation of Kawasaki disease tend to have prolonged hospitalization but short- and long-term coronary outcomes appear to be similar.

Clinical prediction models for young febrile infants at the emergency department: an international validation study

Evelien de Vos-Kerkhof, Borja Gomez, Karen Milcent, Ewout W Steyerberg, Ruud Gerard Nijman, Frank J Smit, Santiago Mintegi, Henriette A Moll, Vincent Gajdos, Rianne Oostenbrink

Arch Dis Child 2018;0:1-9. doi:10.1136/archdischild-2017-314011

Objective: To assess the diagnostic value of existing clinical prediction models (CPM; ie, statistically derived) in febrile young infants at risk for serious bacterial infections.

Methods: A systematic literature review identified eight CPMs for predicting serious bacterial infections in febrile children. We validated these CPMs on four validation cohorts of febrile children in Spain (age <3 months), France (age <3 months) and two cohorts in the Netherlands (age 1-3 months and >3-12 months). We evaluated the performance of the CPMs by sensitivity/specificity, area under the receiver operating characteristic curve (AUC) and calibration studies.

Results: The original cohorts in which the prediction rules were developed (derivation cohorts) ranged from 381 to 15 781 children, with a prevalence of serious

bacterial infections varying from 0.8% to 27% and spanned an age range of 0–16 years. All CPMs originally performed moderately to very well (AUC 0.60-0.93). The four validation cohorts included 159-2204 febrile children, with a median age range of 1.8 (1.2-2.4) months for the three cohorts <3 months and 8.4 (6.0-9.6) months for the cohort >3–12 months of age. The prevalence of serious bacterial infections varied between 15.1% and 17.2% in the three cohorts <3 months and was 9.8% for the cohort >3-12 months of age. Although discriminative values varied greatly, best performance was observed for four CPMs including clinical signs and symptoms, urine dipstick analyses and laboratory markers with AUC ranging from 0.68 to 0.94 in the three cohorts <3 months (ranges sensitivity: 0.48-0.94 and specificity: 0.71-0.97). For the >3-12 months' cohort AUC ranges from 0.80 to 0.89 (ranges sensitivity: 0.70-0.82 and specificity: 0.78-0.90). In general, the specificities exceeded sensitivities in our cohorts, in contrast to derivation cohorts with high sensitivities, although this effect was stronger in infants <3 months than in infants >3-12 months.

Conclusion: We identified four CPMs, including clinical signs and symptoms, urine dipstick analysis and laboratory markers, which can aid clinicians in identifying serious bacterial infections. We suggest clinicians should use CPMs as an adjunctive clinical tool when assessing the risk of serious bacterial infections in febrile young infants.

The relationship between infant iron status and risk of neurological impairment

Buntat, Nurhayati Masloman, Johnny Rompis

Paediatr Indones, November 2017;57: 291-4.

Background: Iron deficiency (ID) is a commonly found nutritional disorder and a persistent problem, especially in Indonesia. Iron deficiency during the critical period in childhood brain development is estimated to cause irreversible damage that hinders infant development.

Objective: To determine the relationship between infant iron status and neurological development.

Methods: We conducted a cross-sectional study at the Growth and Development Outpatient Clinic, Prof. Dr. R. D. Kandou Hospital, Manado, from March to May 2015. By consecutive sampling, we obtained 44 healthy infants aged 7 to 10 months who fulfilled the inclusion criteria. Infants with a history of perinatal complications, such as head trauma, hypoglycemia, respiratory distress syndrome, infection, or malaria were excluded Subjects' serum hemoglobin and ferritin were examined for iron status. Infants' risk of neurological impairment was assessed by the *Bayley Infant Neurodevelopmental Screener* (BINS). Results were analyzed by descriptive analysis for the characteristics and Spearman's rank correlation coefficient analysis for the relationship between iron status and neurological development.

Results: From 14 infants with ID, 8 infants had a high risk of developmental impairment. Of the 30 non-ID subjects, 4 infants had a high risk of developmental impairment. Of the 30 non-ID infants, 16 infants had a low risk of impaired development, while 2 infants with ID had low risk of developmental impairment. Spearman's rho revealed that infant iron deficiency was significantly associated with high risk of neurological impairment. ($r=-0.547$; $P<0.0001$).

Conclusion: Lower serum ferritin levels (iron deficiency) is significantly associated with greater risk of impaired neurological development in infants aged 7-10 months.

DSH NEWS

Honorable Stat Minister, Ministry of Health & Family Welfare Mr. Jahid Malek MP inaugurate Vitamin `A' Plus Campaign on 14 July 2018 at Dhaka Shishu Hospital

Inauguration ceremony of Bangladesh Institute of Child Health (BICH) website by National Professor Shahla Khatun, Chairperson, Management Board of Dhaka Shishu Hospital on 2 October 2018

BICH NEWS

BICH is the academic wing of Dhaka Shishu Hospital. It was established in 30th January, 1983. It is affiliated with Dhaka University, Bangabandhu Sheikh Mujib Medical University (BSMMU) and Bangladesh College of Physicians and Surgeons (BCPS). It has been conducting different courses e.g. DCH, FCPS, MD Paediatrics, MS Paediatric surgery & B.Sc in Health technology. It also conducts different sub-specialty courses e.g. FCPS Neonatology, FCPS Haemato-oncology, FCPS Nephrology, MD Neonatology, MD Haemato-oncology and MD Nephrology. It conducts 3 months certificate course in Paediatrics and 15 days Intensive course for MCPS. It organizes IMCI training and Palli Shishu Rural Health Training. Apart from this, the Institute also runs its regular academic activities. It has established Basic Science Department since 2006.

Diploma course of paediatric nursing has been started from 1st January 2012 and Diploma in paediatric physiotherapy under process.

Library facilities

The library of BICH has a rich collection of updated medical texts and reference books and reputed Medical Journals of home and abroad. BICH has introduced Broad Band facilities which are open to all students, teachers/ consultants of hospital for 24 hours. Facilities of library are also improved by HINARI. Students can download 2230 Medical Journals & more than 50 Paediatric Journals.

Present News

A newly formed classroom in BICH has been named as Prof. Sultan Ahmed Chowdhury as a tribute to First Honarary Director of Dhaka Shishu Hospital.

DS (Child) H J 2018; 34(2): 137

Postgraduate courses/training in paediatrics and child health

1. FCPS in paediatrics : Twice in a year, in the months of January and July.
2. Recognized center by BCPS for training in FCPS (Paeditric surgery) .
3. Recognized centre for course and training in different subspeciality as: Neonatology, Pediatric Nephrology, Paediatric Haematology and Onchology, Paediatric Pulmonology and Paediatric Neuroscience.
3. MD/MS in paediatrics : Part I: In the month of January every year; 2nd and 3rd parts twice every year.
4. DCH course : Once in a year in the month of July.
5. Three months certificate course : The institute every year runs 3 months certificate course on paediatrics for general practitioners & other post graduate candidates e.g. MCPS.
   (Ist August – 31st October)
6. Training programme on IMCI (Integrated management of childhood illness), Essential Newborn Care for doctors and nurses, KMC (Kangaroo Mother Care) traing, ETAT (Emmergency Triage, Assessment and Treatment) training.

Contact Person : Prof. Md. Jahangir Alam
DCH, MCPS, FCPS (Paed)
Editor
Dhaka Shishu (Children) Hospital Journal
Bangladesh Institute of Child Health
Sher-e-Bangla Nagar, Dhaka- 1207.

Contact : Phone No. 9113048, 8122514, PABX: 9104211-20, Ext. 215
E-mail: infodshjournal@gmail.com, profdrmdjahangiralam@gmail.com

DS (Child) H J 2018; 34(2): 138

Students Qualified from Bangladesh Institute of Child Health

Undergoing Courses of BICH

Institution	Courses
Bangabandhu Sheikh Mujib Medical University	MD (Pediatrics) MD Pediatrics Nephrology (sub-specialty) MD Neonatology (sub-specialty) DCH MS (Pediatrics Surgery)
Bangladesh College of Physicians and Surgeons (BCPS)	FCPS Part II (Paediatrics) FCPS Neonatology FCPS Pediatric Nephrology FCPS Hematology & Oncology FCPS Pediatric Surgery FCPS Pediatric Neurology & Development FCPS Pediatric Pulmonology
Dhaka University	B.Sc in Health technology (Lab)
Bangladesh Nursing Council	Diploma in Pediatric Nursing

Student Qualified from BICH till December 2018

Course	Number
DCH	361
MD (Pediatrics)	114
MS (Pediatrics)	103
FCPS (Pediatrics)	28
MD (Neonatology)	13
MD (Pediatrics Nephrology)	5
Total	**624**

Foreign Student Qualified from BICH till January 2013

Country of origin	Course	Number
Nepal	DCH	23
	MS (Ped Surgery)	2
	MD (Ped)	1
India	MD (Ped)	1
Iran	DCH	1
Iraq	DCH	1
Somalia	DCH	1
Sudan	DCH	1
Total		**31**

Present Students (July 2018 to December 2018)

Name of Courses	Number of Students
MD (Pediatrics) Phase - A	14
MD (Neonatology) Phase - A	03
MD (Nephrology) Phase - A	02
MS (Pediatric Surgery) Phase - A	10
FCPS (Pediatrics) Part - II	02
MD (Pediatrics) Part - III	07
MS (Pediatric Surgery)	02
MD (Pediatrics) Part - II	04
DCH (Old)	20
DCH (New)	20
MS (Pediatric Surgery) Part - III	05
Total	89

DS (Child) H J 2018; 34(2): 139

Seminar/Symposium & CME/CPD programs held at BICH (July- December 2018)

Sl. No.	Topic	Unit	Date
1	Update on Neurometabolic Disorder	MU - III	29.07.2018
2	Apnoea in Neonates	MU - IV	05.09.2018
3	Dengue Syndrome		15.09.2018
4	Cystic fibrosis in children	MU - V	14.10.2018
5	Congenital diaphragmatic defects: An update & Recent Guidelines	SU - V	28.10.2018
6	Anthrax: A Hidden Endemic Disease	MU - VI	25.11.2018
7	Poisoning in Paediatrics	MU - VI	23.12.2018

INSTRUCTIONS FOR AUTHORS

Dhaka Shishu Hospital Journal is the official organ of BICH which is the academic wing of DSH. It is published twice a year since 1984. The present editorial board has decided that the cover design will be in accordance with the subject of editorial in each issue. The editor welcome articles to be published in the journal as leading article, original article, review article, case report, current issues of child health, short report and junior's page where trainee doctors are encouraged to publish their topic of interest.

Original papers written in *english* will be considered for publication provided these have not been published previously and are not under consideration for publication elsewhere.

Conditions for manuscript submission:

- All manuscripts will be subjected to peer and editorial review.
- Accepted manuscripts become the property of the *Dhaka Shishu Hospital Journal*. Any reproduction in whole or part will require written permission from the editorial board of the journal.
- The author should obtain written permission from appropriate authority if the manuscript contains any table; data or illustration from previously published in other journals. The letter of permission should be submitted with the manuscript.
- If the photographs are not disguised, permission from the patient or parents/ guardians to print should accompany the manuscript. Otherwise identity will be blackened out.
- Rejected manuscripts/electronic copies/ illustrations/photographs will not be returned to the authors.
- Editors are not responsible for courier/postal failure.

Manuscript preparation: The format of the Dhaka Shishu Hospital Journal complies with *"Uniform Requirements for Manuscripts Submitted to Biomedical Journals"* published by the International Committee of Medical Journal Editors in Vancouver, British Columbia in 1979 (the widely accepted ***"Vancouver style"***)' published in the Annals of Internal Medicine 1982; 96: 766-71. All scientific units should be expressed in *System International (SI) units*. Authors are referred to Annals of Internal Medicine 1987; 106: 114-29 for guidance in the use of SI units. All drugs should be mentioned in their generic form.

- Should be typed in english and on one side of A4 (290 x 210cm) size white paper, using *Times New Roman* font size 12, with single space.
- There should be one original and two paper copies and one IBM compatible electronic copy. (CD or Pen drive)
- There should be a margin of 2.5 cm at top and bottom, and 1.2 cm left and right.
- Pages should be numbered in english numerical at the upper right hand, consecutively, beginning with the title page.
- Manuscripts should be submitted in the following order:
  - Title : should not exceed 100 characters (Font size 16, bold)
  - Name of authors, e.g. 1. Prof. Saiful Islam FCPS, FRCP, 2. Dr. Nurun Nahar MD, these two author's name will be written like this; S Islam[1], N Nahar[2], etc. (Font size 12) Author's designation and name of place of study will be written after the end of the abstract. (Font size 10).
  - ***Abstract with a specific format with five sections (about 350 words maximum): Background, Objective, Methodology, Results, Conclusion, address of correspondence. All these sections will be Times New Roman, Font size 12 and italic , bold but text will not be bold. No references are allowed in the abstract.***

- Text (Introduction, Materials & Methods, Results, Discussion, Conclusion).
- Acknowledgements
- References
- Photographs:
  - In CD/ Pen drive
  - With appropriate labeling (number in English numerical, title of photographs and title of manuscripts.) It should be placed in appropriate place of the article.
- Illustrations:
  - All illustrations should be cited in the text
  - Illustration should be numbered in English numerical and labeled properly, placed appropriately in relation to text of manuscript.
- Tables:
  - Should be appropriately titled.
  - Numbered with Roman numerical serially in order of text description.
  - Abbreviations if used, should be explained in footnotes.
  - Same table should not be repeated as chart.
- Figures:
  - Should be appropriately titled and title will be placed below the figure.
  - Numbered with English numerical serially in order of text description.
- Placement:
  - All photographs, illustrations, tables and figures should be placed in the text in their appropriate places where their description are given.
- References:
  - *References from journal* should be indicated by superscript numbers consecutively in the text and placed after full stop (e.g. ….has been reported.[1] or as shown by Akbar[2] .....) in the order in which they are first mentioned and should be listed in numerical order on a separate sheet at the end of the article.
  - References cited only in tables or legends or illustrations should be numbered in accordance with a sequence established by the first mention in the text.
  - Titles of journals should be abbreviated according to Index Medicus or given in full.
  - References must include: (i) all authors, surnames and initials (if there are 6 authors or fewer) or if there are more than 6 authors, the first six authors followed by et al; (ii) the full title of the paper; (iii) the abbreviated or full title of the journal in italic; (iv) the year of publication; (v) the volume no will be bold; (vi) the first and last page numbers followed by full stop. *Example*: Khan NZ. A study of mentally handicapped children: aetiology and associated factors. *Bangladesh Journal of Child Health* 1985; **9**(2):102-08.
  - *References from books* must include: (i) authors name, (ii) title of article, (iii) editors name/s, (iv) name of the chapter, (v) place of publication, (vi) name of publisher, (vii) year of publication and page numbers. *Example*: Razvani I. An approach to inborn errors of metabolism. In: Behrman RE, Kliegman RM, Jenson HB, editors. Nelson Textbook of Paediatrics. Philadelphia: Saunders, 2004: p.397-98.
  - *Documents in electronic format* must include: (i) title, (ii) authors name, (iii) year of publication, (iv) web site address, (v) date of access. *Example*: United Nations Programme on HIV/AIDS. Children living in a world with AIDS. Geneva, 1978 (htpp://www.....), accessed on (dd/mm/year).

Manuscripts Submission: the manuscripts should be submitted to the editor/ associate editors with a covering letter, mentioning that the work has not been published or submitted for publication anywhere else.

Reprints for the authors: 2 copies of original journal and five copies of each article will be provided to the corresponding author free of cost.

Copy right: No part of the materials published in this journal may be reproduced, stored or transmitted without prior written permission of the editorial board.