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Editorial

Advances in technology and training in paediatric cardiology have improved long-term outcome and promised better quality of life. With facilities for accurate diagnosis and scope of complete correction, more and more children are undergoing cardiac intervention and surgical treatment for congenital heart diseases. In every edition Paediatric Heart Journal of Bangladesh, we try to improve it with recent knowledge and development in the field of paediatric cardiology. This edition of our journal contains a number of articles which are worth appreciable.

It is important to identify and evaluate strategies to enhance early detection as timely recognition of Critical CHD could improve outcomes. Pulse oximetry is effective in identifying infants with CCHD and is much less expensive than screening all newborns with echocardiography. Hemodynamics are variable between individual babies and also within the same baby with time. There is a wealth of hemodynamic information that can be derived by functional echocardiography in neonatal. The decision of treatment of PDA should be individualized, according to clinical, echocardiographic, and biochemical parameters that validate hemodynamic significance of PDA. Some of these controversies in neonatal cardiology is discussed in the leading article.

During last few decades outcome of Transposition of the great arteries (TGA) management has improved significantly in western world and few of our neighboring countries as well. Since the first successful arterial switch operation (ASO) in 1975, survival rates have increased with refinement of surgical techniques and improved medical management. In many countries with emerging economics, management of TGA is still challenging. Although ASO is the ideal surgical management option for D-TGA, in the countries with emerging economics patients sometimes present late who are not suitable for ASO. In our country despite having huge number of patient burden, only one center is doing limited number of surgical management of TGA. In this issue experience of NHFH&RI regarding modalities of surgical treatment, document the immediate outcome, demonstrate major post-operative events of TGA patients after ASO is shared.

There is increasing demand for dedicated personnel for the specialized intensive care of critically ill children suffering from cardiac diseases. A dedicated team dictating specialized intensive care has translated into better outcomes in several centers. Over recent decades, specialized pediatric cardiac intensive care has emerged as a central component in the management of critically ill neonatal and pediatric patients with congenital and acquired heart disease worldwide. The majority of developed centers have dedicated pediatric cardiac intensive care units to care for pediatric cardiac patients. In developing nations with limited resources, pediatric cardiac intensive care is yet to take root as a distinctive discipline In this issue experience of Paediatric Cardiac Intensive Care Unit of Dhaka Shishu (Children) Hospital is shared.

Prevalence of hypertension in children has increased significantly in recent times, in part related to the epidemic of childhood obesity. Identification and treatment of hypertension in childhood is likely to favorably impact on cardiovascular disease in adulthood. Identification of hypertensive children continues to be problematic because of incomplete blood pressure screening during routine pediatric clinical visits. Once diagnosed as definitive hypertension, the causes of secondary hypertension should be determined and appropriately treated. In children with primary hypertension, a combination of life-style changes (diet and exercise) and drug therapy should be instituted depending upon the stage of the hypertension.

These are some of the topics discussed in this issue of Paediatric Heart Journal of Bangladesh. Any comments or criticism will be highly appreciated.

Editor

Leading Article

Issues and Controversies in Neonatal Cardiology

Manzoor Hussain

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Introduction

The care of Congenial Heart Disease (CHD) has improved for the last 60 years.[1] With developments in early diagnosis and rapid stabilization, improvements in imaging, advanced interventional techniques, and newer treatment options for pulmonary artery hypertension and heart failure have complemented the growth in this field. Diagnosis of CHD during neonatal period is increasing day by day. Recent hospital based data revealed that 30% CHD is detected during neonatal period in Bangladesh.[2] If these affected neonates left untreated causes serious morbidity and mortality, therefore early detection and proper intervention is very important. The field of Neonatal cardiology is full of challenges and controversies.

What goes wrong at the first step of diagnosis: Detection of CHD at birth and early infancy is difficult due to limited access to health care and limited exposure of most physicians to pediatric cardiology.

Importance of early identification: Like other developing countries, Bangladesh is facing a multitude of health problems and CHD is one of them. Many of the CHDs are critical congenital heart disease (CCHD) which requires surgery or catheter intervention in the early period of life and responsible for more deaths than any other type of malformation.[3] Without early recognition, diagnosis and treatment, a majority of infants with CHD die in their first month of life in developing countries.[4] Too often referrals are delayed because of ignorance or limited knowledge of CHD or its natural history, inaccurate diagnosis and lack of awareness about available facilities. As the majority deaths of infants with CHD occur during the first month, with early diagnosis most of these babies could be saved.

Prenatal diagnosis: Over the past two decades, imaging has become the principle diagnostic tool in prenatal detection of foetal malformations. Antenatal diagnosis of CHD by doing fetal echocardiography has been shown to benefit outcome.[5] Generally, detailed fetal echocardiography is not performed routinely for prenatal screening but is reserved for cases which are at high-risk for CHD. Routine fetal echocardiography is recommended when there is risk factors like family history of CHD; coexisting maternal disease; exposure to teratogens in early pregnancy; infections such as parvovirus B19, rubella, coxsackie; abnormal karyotype and extracardiac fetal anomalies such as diaphragmatic hernia, exomphalos noted on a general fetal sonogram. But only 10 percent of the fetuses with cardiac anomalies have identified risk factors. In such cases, detailed fetal echocardiography is commonly done between 18 and 22 weeks of gestation.[6] Appropriate detection of cardiac abnormalities can only be achieved by carrying out routine fetal echocardiography in all fetuses in the second trimester, irrespective of the presence or absence of risk factors for the development of CHD. This is not yet possible in a developing country like Bangladesh.[7]

Pulse oximetry screening for CHD: Pulse oximetry has the potential to identify hypoxemia that might not otherwise produce visible cyanosis. Pulse oximetry is used routinely in the assessment of neonate in

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neonatal intensive care units and emergency departments. It has been proposed as an adjunct to the assessment of the newborn in the delivery room.[8] Critical CHD in the newborn may have borderline low oxygen saturation with unrecognized cyanosis clinically. Pulse oximetry is highly specific for detection of critical congenital heart defects with moderate sensitivity that meets criteria for universal screening.[9] Arterial oxygen saturation varies considerably in the first 24 hours, with many healthy newborns having arterial saturations of less than 95%. As such, oximetry screening before 24 hours of life can result in a significant number of false-positive results due to transition from fetal to neonatal circulation. A study from the United Kingdom reported that the false-positive rate was as high as 5% when oximetry screening was performed in the first 24 hours compared with 1% at the time of hospital discharge.[10] Later screening can miss an opportunity for intervention for defects that are impacted by closing PDA. It should be done at 24-48 hours of life as a screening method to detect Critical CHD.

Clinical examination still the key: A normal examination of newborn does not exclude heart disease. The clinician must remain vigilant to detect early signs and refer infants promptly to a pediatric cardiologist for definitive evaluation.[11] Sensitivity of clinical assessment by cardiologist is found 89.8% and specificity 75%.[12] So clinical examination still the key to identify CHD.

Urgent referral: High index of suspicion and astute acumen are essential to decision making. When PDA is opened widely, many serious malformations may not be noticed easily in the early life. They would progress as severe acidosis, shock, cyanosis or even death as PDA constricts after few hours to days. Recent studies showed that a significant proportion of neonate with critical CHD experienced late or no referral to cardiac specialty center resulting in significant number of death.[13] Factors like, late presentation of cases, associated co-morbid conditions, understaffing of units and limited resources contribute to suboptimal outcome in those who undergo corrective surgery.[14]

Vital signs monitoring in neonate: How much we can rely? Newborn may have a variety of haemodynamic problems with a variable and complex pathophysiology, that at times have limited clinical manifestations. This cardiovascular vulnerability stems from particular features of the newborn, such as incomplete myocardial development, presence of fetal shunts, changes in systemic and pulmonary vascular resistance and complex hemodynamic changes that take place during transition to extrauterine life. Despite progressive technological advances in neonatal intensive care, haemodynamic monitoring in newborns is still based on assessment of continuous heart rate, blood pressure (BP), acid base status, urine output and capillary refill time.[15] These measures provide important and useful information to the clinician. Clinical assessment and the classic parameters mentioned before to elucidate the underlying pathophysiology in haemodynamic disturbances can sometime lead to incorrect assumptions and therapeutic decisions, which may even cause harm.[16]

Heart rate: Rising pulse rate in adult is indicative of hypovolemia which relies on mature autonomic nervous system as an attempt to sustain CO. Increase in HR is the most effective way to increase CO. So tachycardia is a reliable sign of hypotension and circulatory inadequacy. But if myocardial damage has occurred it produces hypotension, and neonate failed to maintain a sustained increase HR. Besides this neonate with hypotension also may be hypoxic and hypoxia vagally mediate lowering HR. In preterm neonate systemic blood flow and HR are not significantly correlated due to immature myocardium and autonomic nervous system.[15]

Capillary refill time (CRT): CRT >3 sec is a reliable sign of shock in neonate and it has 55% sensitivity and 81% specificity in predicting low systemic pressure (systemic blood flow measured by superior vena caval flow).[17] But it can be influenced by site, pressure time, temperature, flow mechanis and drugs.[18]

Blood pressure (BP): BP is widely accepted parameter in assessing cardiovascular

stability. Neonatal BP is considered to be adequate as long as urine output (>1ml/kg/hr) and CRT (<3 sec) are within normal limit. A direct reading from indwelling arterial catheter represents gold standard. Correlation between invasive and noninvasive measurement of BP is generally good. Indirect determination is higher than these obtained directly by 3-5 mm Hg. But noninvasive BP in preterm is problematic as it is more dependent on appropriate cuff size.[19]

Urine output: The immature renal tubule in VLBW infants is inefficient at concentrating the urine and therefore may be unable to appropriately reduce urine flow. Even if the GFR is decreased markedly, there may be little or no change in urine output.[20]

Situations in which serial assessment is needed to evaluate response to treatment or the hemodynamic changes that take place during transition from fetal to postnatal circulation, it is important to consider that these changes can be very different from what might be assumed clinically based on conventional assessment.[15] These variables associated in varying degrees to tissue perfusion, which is the key haemodynamic parameter, and for which an adequate monitoring method has yet to be developed.[21]

Functional Echo in NICU: Is it necessary? The term functional echocardiography have been introduced to describe the echocardiography as an adjunct in clinical assessment of the hemodynamic status in neonate. It provides noninvasive estimates of a wide range of hemodynamic parameters. Infants without any clinical suspicion of CHD the first echocardiographic study must be a comprehensive examination assessing structure and function.[15] There is a wealth of hemodynamic information that can be derived by functional echocardiography. Hemodynamics are variable between individual babies and also within the same baby with time. Without echocardiography there will be guessing the hemodynamics which will be wrong most of the time. It need to be available at any time and in short notice in NICU. Neonatologists themselves have to develop echocardiographic skills in close collaboration with their cardiologist colleague.

Controversies in the treatment of PDA in preterm infants: Whether or not to use prophylaxis, when to treat a moderate-to-large PDA, enteral feeding during PDA treatment and when to do surgical closure are the controversies regarding PDA. Current controversy of treatment is due to high likelihood of spontaneous ductus closure and the absence of RCTs. No specific clinical or echocardiographic criteria have been developed on which treatment of PDA could be based. In recent years there has been a growing debate about whether or not to treat a persistent PDA in neonates. The preterm PDA has shifted from being viewed as a pathologic condition causing morbidities and mortality in the preterm infant to being proposed as an innocent physiological bystander.[22] Echocardiography is widely used to define haemodynamically significant PDA requiring treatment and to exclude duct-dependent congenital heart disease. Although there are no stringent echocardiographic criteria to define the need for therapeutic intervention, several echocardiographic parameters have been correlated with PDA hemodynamic significance and with therapeutic responsiveness. These parameters include: PDA diameter >1.4 mm, the internal diameter of the ductus/body surface area ratio, a low-velocity pulsatile flow pattern, left atrium to aorta ratio >1.4, and diastolic reverse flow in the aorta, mesenteric, cerebral, and renal arteries.[23,24] A conservative approach to the treatment of PDA involves fluid restriction and "watchful waiting." Diuretics lack evidence justifying routine use, but they may be useful if the neonate is exhibiting signs of CHF while waiting for spontaneous closure of the PDA.[25] Inhibition of prostaglandin synthesis with nonselective inhibitors of cyclooxygenase - 1 and - 2 (e.g., indomethacin and ibuprofen) appears to be an effective alternative to surgical ligation.[26] Recently, paracetamol has been shown to be an alternative treatment for closure of PDA because of its safety profile and low cost.[27] Prophylactic treatment approach has several important short-term benefits but it results in over-treatment of infants that might close their ductus spontaneously.[28] The use of

surgical ligation as a first line of treatment for PDA is influenced mainly by surgical availability. There is no evidence to support surgery as the preferable treatment approach. Studies found that infants randomized to surgery had higher rates of pneumothorax and retinopathy of prematurity but no difference in other outcomes, including mortality.[29] There were no significant differences in the rates of BPD, sepsis, ROP, neurologic injury or mortality between: aggressive approach using early surgical ligation (within 2 days) when the infant's PDA failed to close after indomethacin treatment and the conservative approach continued feedings in the presence of a PDA and only ligated the PDA if cardiopulmonary compromise developed.[30] Empirical use of medication to close the duct without doing echocardiography is not recommended.

Use of anti failure drug in neonate with CHD: The current maintenance treatment for young infants with CHD with heart failure remains controversial. To a large extent, it is based on extrapolation of data derived from trials in adult populations. There are only a few randomized trials in Pediatrics.[31] The goals of therapy are to maintain circulatory and end-organ function and to allow for recovery and reverse remodeling to occur. A category of heart failure unique to pediatrics is volume overload lesions typically left-to-right shunt lesions. The RV is often abnormal in CHD and can fail as a result of volume and pressure overload in a variety of malformations.[32] The goals of therapy are to maintain circulatory and end-organ function and to allow for recovery and reverse remodeling to occur.

Digoxin: Historically digoxin is the mainstay of medical therapy of heart failure in children. It is indicated in heart failure associated with reduced systolic function of heart. Its role in heart failure secondary to left to right shunt lesions with normal systolic function is contradictory.[33]

Frusemide: Frusemide is proven to be beneficial for symptomatic relief in heart failure. No survival benefit has been shown for patients with heart failure.[34] Frusemide continues to be used in volume-overload conditions to decrease pulmonary congestion and thus decrease the work of breathing.[35]

Spironolactone: Spironolactone has beneficial effects on cardiac remodeling. For patients taking more than 2 mg/kg of oral furosemide daily without ACE inhibitors, spironolactone should be added for its potassium-sparing effect.[34]

ACE inhibitors: Afterload reduction with ACEi is indicated in patients who have large left-to-right shunts at the ventricular or arterial level, left-sided regurgitant lesions, or poor systolic function.[36] Currently ACEi therapy is recommended as the first line treatment for heart failure in children, when it is not secondary to an obstructive lesion. In neonate it should be used very cautiously due to concerning incidence of renal failure.[34]

Beta blocker: Exact indications and benefits of beta blocker (carvedilol) therapy in children with heart failure remain somewhat unclear.[37]

Issues to be considered with the use of anti failure drug: Antifilure drugs should be continued to minimize symptoms and optimize growth until a definitive procedure can be performed. The drugs should be stopped during diarrhoeal episodes.

Conclusion

It is important to identify and evaluate strategies to enhance early detection as timely recognition of CHD could improve outcomes. Pulse oximetry is effective in identifying infants with CCHD and is much less expensive than screening all newborns with echocardiography. Hemodynamics are variable between individual babies and also within the same baby with time. There is a wealth of hemodynamic information that can be derived by functional echocardiography. The decision of treatment of PDA should be individualized, according to clinical, echocardiographic, and biochemical parameters that validate hemodynamic significance of PDA. As the available evidence does not support prophylactic or presymptomatic approach, expectant symptomatic treatment for hemodynamically significant PDA seems to be the most reasonable approach. Empirical use of

medication to close the duct without doing echocardiography is not recommended. The current maintenance treatment for CHD with heart failure remains controversial. The goals of therapy are to maintain circulatory and end-organ function and to allow for recovery and reverse remodeling to occur.

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Original Article

Clinical Profile & Disease Pattern of Admission in Pediatric Cardiac Intensive Care Unit of Dhaka Shishu (Children) Hospital

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Abstract

Background: *Intensive care is predominantly concerned with the management of patients with acute life threatening conditions in a specialized unit. Children having acute cardiovascular compromise required more meticulous care in pediatric cardiac intensive care unit.*

Objective: *This study was conducted to see the clinical profile and disease pattern among patients in Pediatric Cardiac Intensive Care.*

Methodology: *This cross sectional study was conducted in Pediatric Cardiac Intensive Care Unit (PCICU) of Dhaka Shishu (Children) Hospital from February 2013 to January 2014. On admission, a detailed history of the illness was taken, thorough general and systemic examinations and findings of the performed investigations, relevant associated medical conditions was recorded carefully and during treatment complications were recorded from patients' file. Data were processed and analyzed using computer software SPSS version 21.*

Results: *Total 289 children with heart disease for cardiac medical care and 37 children for post-surgical care were admitted during study period. Among the medical cases 40.83% were neonate, 33.21% were 1 month-1 yr, 25.94% were >1 yr. Male were 62.28% and female were 37.71% with a male female ratio 1.65:1. A large group of children admitted with heart failure (16.60%), pneumonia (14.50%), sepsis (13.10%) & metabolic acidosis (12.10%). Majority of CHD were of Acyanotic CHD (74.0%). Among acyanotic CHD majority were of VSD (22.2%) & among cyanotic CHD majority are of TOF (24.6%). Among the admitted patients 62.62% was discharged, 24.56% died, 9.68% left against medical advice and 3% referred to other specialized cardiac centre or abroad. Mean age of children who underwent cardiac surgery was 4.97±3.6 (range 1.1-14 years), weight 14.13±7.7 (range 4.9-41 Kg) and mean post-operative stay was 5.18±4.59 days (range 1-19 days). Three (8.1%) cases died after cardiac surgery. Outcome of ASD and VSD closure, PDA ligation was excellent. Among total correction of TOF 75% survived.*

Conclusion: *Pediatric cardiac intensive care in developing nations is still in infancy. As a newly established center survival of cardiac intervention, pediatric cardiac surgery and those who admitted with medical care is satisfactory.*

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Introduction

The practice of pediatric cardiac intensive care depends on a collaborative effort from all disciplines involved in the care of critically ill pediatric patients with cardiovascular disease. Pediatric cardiac intensive care has evolved as a distinct limb of efficient pediatric cardiac programs in the developed nations. With increasing demand for congenital heart surgery in the developing nations, concept of pediatric cardiac intensive care units (PCICU) is critical to the success.[1] In adults, cardiac disease is not uncommon and the majority of causes of sudden cardiac death are due to atherosclerotic coronary artery disease.[2] Cardiac death in children is a rare occurrence and the cause differs based on the age of the child. Disease pattern in pediatric intensive care unit particularly in early age group is a sensitive indicator of the availability, utilization and effectiveness of mother and child health services. However, disease pattern changes between different places and time to time even at the same place.[3] Most common congenital heart lesions were ASD (26%), VSD (16.9%), PDA (18%), Tetralogy of Fallot (14%), Pulmonary stenosis (7.75%) etc.[4]

In a recent retrospective search of a computerized database and medical case notes for all acute cardiac admissions to PICU by Giridhar et al[5] found that new cardiac diagnoses presenting to a with obstructive left heart lesions, transposition of the great arteries (TGA), total anomalous pulmonary venous drainage (TAPVD), dilated cardiomyopathy, arrhythmia and others. Previous research also investigating of 100 consecutive infants of diabetic mothers (IDMs) at King Khalid University Hospital in Riyadh reported most common echocardiographic findings were patent ductus arteriosus, patent foramen ovale, atrial septal defect, small muscular ventricular septal defect, mitral valve prolapse and pulmonary stenosis. Severe forms of CHD encountered were D-transposition of great arteries, tetralogy of Fallot and hypoplastic left heart syndrome.[6]

The pediatric population in our country have a significantly higher incidence and prevalence of serious cardiac diseases. This is contributed to by a variety of factors that include a lack of early correction of congenital cardiac abnormalities that results in accumulation of a large number of children with uncorrected anomalies who survive the neonatal period. In addition, many of these children develop more rapid deterioration in their clinical condition as a result of accelerated forms of secondary changes in the heart and other organs. Some of these changes could be the result of genetic and/or environmental factors such as pollution or infection.[7] Cardiovascular compromise due to previously unrecognized congenital or acquired heart disease is associated with clinically significant morbidity and mortality. Patients are admitted to a pediatric cardiac intensive care unit require a very high level of monitoring of vital signs and other body functions and provide the exact estimate of the burden.[8] Pediatric cardiac intensive care is still in the evolving phase in many emerging nations, including Bangladesh. However, data specific to dedicated cardiac ICUs are relatively few. So this study was undertaken to see the clinical profile and disease pattern among patients in Pediatric Cardiac Intensive Care unit of Dhaka Shishu (Children) Hospital.

Materials and Methods

The study was conducted through a cross sectional observational approach at Pediatric Cardiac Intensive Care Unit (PCICU) of Dhaka Shishu (Children) Hospital from February 2013 to January 2014. All the patients with CHD admitted in PCICU and patients with CHD transferred to PCICU for post-operative care during the data collection period were included. On admission, a detailed history of the illness of traditional and emergent cardiovascular disorders was taken from the patient, thorough general & systemic examinations and findings of the performed investigations, relevant associated medical conditions will be recorded carefully and during treatment complications were recorded. Data were collected and after proper cleaning data were analyzed thoroughly. Data entry and analysis was done by using

SPSS version 21. Written consent was taken from all enrolled patient's parents or attendants and permission was also taken from ethical review committee of Bangladesh Institute of Child Health and Dhaka Shishu (Children) Hospital.

Result

Total 289 children were admitted in PCICU, among them 118 (40.83%) were neonate, 96 (33.21%) were 1 month - 1 yr, 58 (20.06%) were 1 yr-5yr, 17 (5.88%) were > 5 yr old (Table-I).

Table-I

Distribution of study population by age (n=289)

Age	Frequency	Percent
Neonate	118	40.83%
1 month- 1 year	96	33.21%
1-5 Year	58	20.06%
>5 years	17	5.88%

Among the neonates male were 62.28% and female were 37.71% with a male female ratio 1.65:1 (Fig.-1).

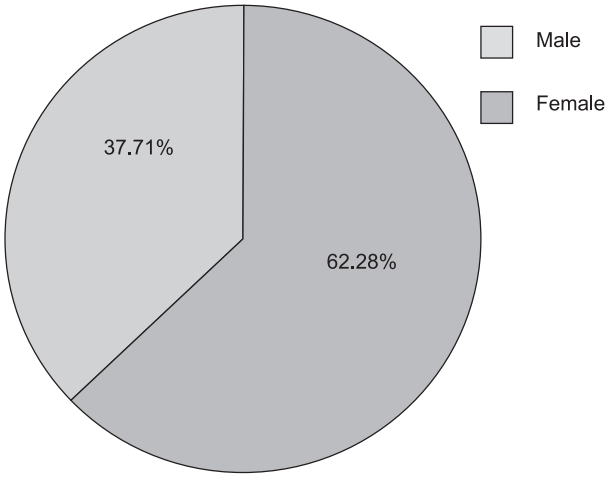

Fig.-1: *Sex distribution among the admitted children (n=289)*

A large group of children admitted with heart failure (16.60%), pneumonia (14.50%), sepsis (13.10%) & metabolic acidosis (12.10%) [Table-II].

Table-II

Presenting problems during admission in PCICU

Admitting Problem	Number (%)
Heart Failure	48 (16.6%)
Pneumonia	42 (14.5%)
Cyanotic Spell	20 (6.92%)
Shock	28 (9.68%)
Sepsis	38 (13.1%)
Metabolic Acidosis	35(12.1%)
Respiratory Acidosis	30 (10.3%)
Renal Failure	12 (4.15%)
Arrhythmia	06 (2.07%)
Anemia	15 (5.19%)
Duct dependent pulmonary circulation	10 (3.46%)
Duct dependent systemic circulation	05 (1.73%)

Majority of admitted children are of Congenital CHD, 275 out of 289 (95.1%) and only 14 (4.9%) is acquired (Table-III).

Table-III

Distribution of CHD among study population (n=289)

Type of heart disease		Number	Percent
Congenital	Acyanotic	202	69.9
	Cyanotic	73	25.2
Total		275	95.1
Acquired	SVT	3	1.1
	Dilated Cardiomyopathy	7	2.4
	Rheumatic heart disease	2	0.7
	Infective Endocarditis	2	0.7
Total		14	4.9

Among acyanotic CHD majority are of VSD (22.20%), followed by ASD (18.80%) and PDA (12.90%) [Fig.-2].

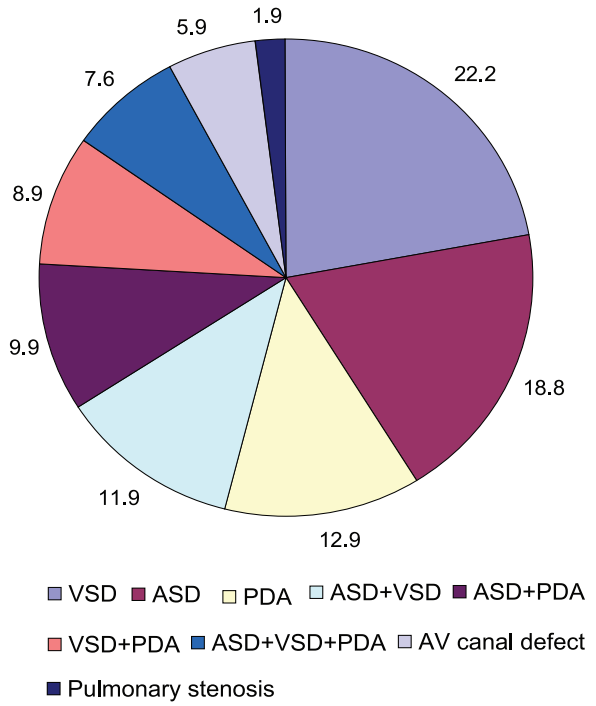

Fig.-2: *Pattern of Acyanotic CHD (n=202)*

Among cyanotic CHD majority are of TºF (24.60%), followed by D-TGA (21.90%) and Pulmonary Atresia (13.60%) [Table-IV].

Table-IV
Distribution of cyanotic CHD among study population (n=73)

Cyanotic CHD	Number (%)
Tetralogy of Fallot	18 (24.60%)
D-Transposition of great arteries	16 (21.90%)
Tricuspid Atresia with Shunt	08 (10.9%)
Pulmonary Atresia with Shunt	10 (13.60%)
TAPVC with Shunt	08 (10.9%)
Truncus Arteriosus with Shunt	03 (4.10%)
Single Ventricle	05 (6.84%)
Coarctation of Aorta with Shunt	03 (4.10%)
Hypoplastic Left Heart Syndrome	02 (2.73%)

Prostaglandin infusion was required in 10 cases followed by BAS in 3 cases. SVT was managed with adenosine in 3 cases, CV line was given in 10, pericardiocentesis in 3, cyanotic spell management in 20 and ventilator care in 30 cases. Successful therapeutic cardiac intervention done among 06 patient during this period (Table-V).

Table-V
Distribution of special procedures done in PCICU

Special interventions	Number
Prostaglandin infusion	10
BAS	3
SVT management with adenosine	3
CV line	10
Ventilator care	30
Pericardiocentesis	3
Management of Cyanotic spell	20
PDA devise closure	02
ASD devise closure	02
BPV for severe pulmonary stenosis	01
Balloon dilatation of CoA	01

Among the admitted patients 181 (62.62%) was discharged, 71 (24.56%) died, 28 (9.68%) leave against medical advice and 09 (3%) referred to other specialized pediatric cardiac centre of home & abroad (Fig.-3).

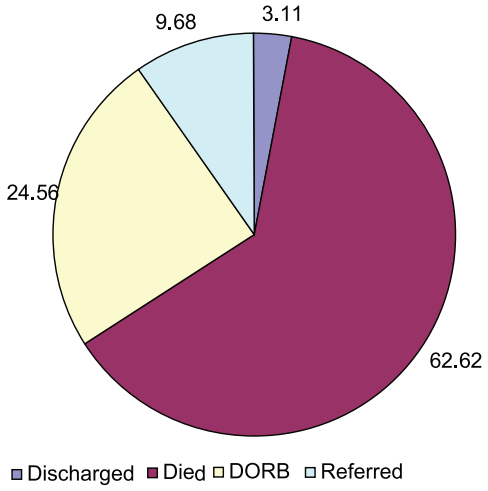

Fig.-3: *Distribution of outcome of patients in PCICU (n=289)*

Trends of survival improving gradually 12 out of 24 (50%) on Feb,2013 & 20 out of 26 (76.9%) on Jan,2014 and mortality decreasing, 8 out of 24(33.33%) on Feb,13 & 4 out of 26 (15.38%) on Jan,14 (Fig.-4).

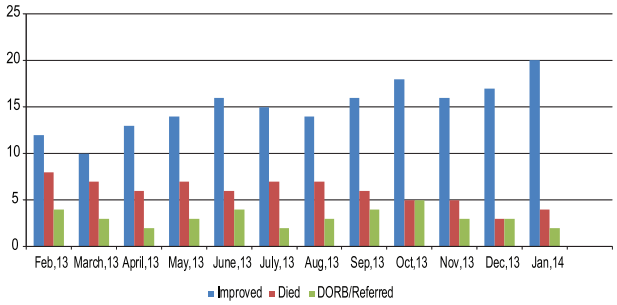

Fig.-4: *Monthly trends of outcome of admitted children (n=289)*

Total 37 cardiac surgery was done during the study period and mean age was 4.97±3.6 (1.1-14 years), mean weight 14.13±7.7 (range 4.9-41 Kg) and mean post-operative stay was 5.18±4.59 days (range 1-19 days) [Table-VI].

Table-VI

Distribution age, weight and PCICU stay of cardiac surgery cases (n=37)

	Mean±SD (Range)
Age (Years)	4.97±3.6 (1.1-14)
Weight (Kg)	14.13±7.7 (4.9-41)
Hospital stay (Days)	5.18±4.59 (1-19)

Only 03(8.1%) cases died after cardiac surgery. Outcome of ASD, VSD closure, PDA ligation was excellent. Among total correction of TOF 75% survived (Table-VII).

Table-VII

Outcome of cardiac surgery cases (n=37)

CHD	Survived(%)	Died(%)
PDA ligation	11(100)	-
VSD closure	6(86)	1(14)
ASD closure	4(100)	-
VSD, ASD closure with PDA ligation	1(100)	-
RVOT & PA Augmentation	2(100)	-
Repair of PAPVC with ASD	1(100)	-
TOF correction	6(75)	2(25)
VSD with ASD closure	3(100)	-

Discussion

There is wide gap between the developed and developing countries regarding pediatric cardiac care. Absence of pediatric cardiac centers, presence of cardiac centers only in large cities, unstable political systems affecting social stability, and absence of specific health care policies in various countries are the reasons for this variation.[9] Although there has been establishment of tertiary-level pediatric cardiac care services in public hospitals, the time has arrived to further enhancement in the country. Due to a lack of resources cardiac care inadequacy cannot be solved within a short span of time. Giving urgent attention to 2 important reasons for inadequate pediatric cardiac treatment in Bangladesh can change the scenario drastically. These are (1) lack of awareness about CHD and (2) high rate of unsupervised home deliveries because of which CHD are not detected at birth.

In developing nations with limited infra-structure, human, and material resources, pediatric cardiac intensive care is yet to take roots as a distinctive discipline. As a result, many models exist. Pediatric heart programs are often attached to well-established adult cardiology and cardiac surgery programs, and PCICU care is sometimes delivered in a common setting with shared space, infrastructure, and personnel. In small private establishments, it is delivered by a small group of professionals attached to the surgical unit.[10,11] In a recent survey of various pediatric cardiac intensive care programs in the United States and Europe, 19 (35%) institutions had a dedicated cardiac ICU, whereas, in 35 hospitals (65%) pediatric cardiac patients were cared for within a general pediatric cardiac intensive care unit.[12] A retrospective study was conducted to see whether there is there a difference between the pediatric intensive care unit and the PCICU on clinical outcome measures of pediatric cardiac postoperative patients and nursing resources and found that majority of quality measures were not statistically different between the 2 ICUs.[13] One study evaluated the benefits of a dedicated Pediatric Cardiac Intensive Care Unit (PCICU) in the early

postoperative outcomes of patients undergoing surgery for congenital heart disease and found that establishment of a dedicated pediatric cardiac intensive care unit has shown better outcomes in terms of earlier extubation, de-intensification, and discharge from the ICU.[14] Studies found that dedicated intensive care units (ICU), run by an intensivist, who led a multiprofessional team is known to produce better outcomes.[15,16] This study was conducted among both pediatric cardiac medical cases and post-operative cases. Among the admitted patients 62.62% was discharged, 24.56% died, 9.68% leave against medical advice and 3% referred to home or abroad. Among post-surgical cases mortality was 8.1%. Mattos et al[17] found 14.7% mortality among post-surgical cases. One previous Bangladeshi study showed that case fatality rate was 15.9% in 2007, 8.3% in 2008, 12% in 2009 and came down to 8.75% in 2010.[18] Complicated cardiac surgeries are not yet started in this center and it may be the cause of lower mortality in the present study.

The distribution of specific lesions was different between the live births and stillbirths. In China among the live births, the top three lesions were ventricular septal defect (VSD), patent ductus arteriosus, and atrial septal defect, which accounted for 34.0%, 23.7%, and 10.8%, respectively.[19] In Bangladesh one multi-center study and one past and present situation analysis found that VSD remains at the top of the list followed by ASD, PDA, TOF and TGA. VSD was the commonest among acyanotic CHD and TOF was the commonest cyanotic CHD.[20,21] This study showed that majority of CHD are of Acyanotic CHD (74.0%). Among acyanotic CHD majority are of VSD (22.20%), followed by ASD (18.80%) and PDA (12.90%) and among cyanotic CHD majority are of TOF (24.60%), followed by D-TGA (21.90%) and Pulmonary Atresia (13.60%).

Hussain et al[20] found that majority cases of CHD were diagnosed during infancy and about 30% were detected during neonatal period. This study showed that 40.83% of PCICU admitted cases were neonate. Pediatric cardiac care in Bangladesh is still in its infancy and in the way of expansion.[22] With the advancement of time early detection rate is gradually increasing in Bangladesh and more CHD are seeking medical treatment early.

In the developed world CCHD can be treated with 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.[23] This study showed that prostaglandin infusion was started in 10 cases of TGA and among them balloon atrial septostomy was done in 3 cases. All the cases were successfully prepared for corrective surgery. Successful therapeutic cardiac intervention done among 06 patient during this period. Outcome of ASD, VSD closure & PDA ligation was excellent. Among total correction of TOF 75% survived.

Conclusions

Pediatric cardiac intensive care in developing nations is still in infancy. As a newly established pediatric cardiac intensive care center survival of pediatric cardiac intervention, surgery and those who admitted with medical care is satisfactory.

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Surgical Management of TGA at National Heart Foundation Hospital: Nouveau Espoir

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Abstract

Background: *Transposition of the great arteries (TGA) is a complex congenital heart disease being most common congenital cyanotic heart disease in infancy. During last few decades outcome of TGA management has improved significantly in western world and few of our neighboring countries as well. In many countries with emerging economics, management of TGA is still challenging.*

Objectives: *To analyze outcome of TGA management in a Tertiary Care Pediatric Cardiac Centre in a resource limited setup of a country from emerging economy.*

Methodology: *This retrospective study was conducted at Pediatric & Congenital Cardiac Program of National Heart Foundation Hospital and Research Institute, Dhaka, Bangladesh over a period of 2 years from July 2014 to June 2016. Data were obtained from departmental Congenital Heart Surgery Database. All patients admitted with a diagnosis of TGA were included in the study. Major preoperative events, surgical procedures, major post operative events and outcomes were recorded. Standard statistical software and tests were used for data analysis.*

Results: *During 2 years period from July 2014 to June 2016 total 26 patients were admitted with a diagnosis of TGA with a median age of 2.3 months. Three (11.5%) patients required preoperative stabilization with Balloon Atrial Septostomy (BAS). Types of procedures include Arterial Switch Operation (ASO) 10 (38.5%), ASO with VSD repair 10 (38.5%), Senning procedure 4 (15.4%), REV procedure 1 (3.8%) and Palliative procedure (Modified Blalock Taussig Shunt) 1 (3.8%). Major post operative events include open sternum 9 (34.6%), re-exploration 5 (19.2%), diaphragmatic palsy 3 (11.5%), Acute Kidney Injury (AKI) requiring dialysis 3 (11.5%), Low Cardiac Output Syndrome (LCOS) 3 (11.5%) and pulmonary hemorrhage due to multiple collaterals 1 (3.8%) case. Median post operative ICU stay was 12 days and mortality 6 (23.1%) cases. Common causes of mortality include LCOS and septicemia.*

Conclusion: *Although reasonably better outcome in the management of TGA has been achieved, still there are scopes of further improvement to achieve better level.*

Key words: *Transposition of Great Arteries (TGA), surgical management, arterial switch operation (ASO).*

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Introduction

TGA is a severe cardiac malformation in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. An alternative terminology describes this malformation as a cardiac anomaly with the combination of concordant atrio-ventricular and discordant ventriculo-arterial connections.[1] TGA is a lethal and relatively frequent malformation accounting for 5-7% of all congenital cardiac malformations.[2] Without intervention, babies born with TGA are doomed to a rapid death.[3]

Since Jatene et al[4] performed the first successful ASO in 1975, survival rates have increased with refinement of surgical techniques and improved medical management. Currently, most treated patients live to adulthood, with a 20-year survival rate of nearly 90%.[5] Strong predictors for poor overall free-reoperation survival are complex TGA, VSD, coronary anomalies, aortic coarctation and Left ventricular outflow treat (LVOT) obstruction or moderate Pulmonary artery stenosis.[6]

Although ASO is the ideal surgical management option for D-TGA, in the countries with emerging economics patients sometimes present late who are not suitable for ASO.[7] In those cases options are either rapid two staged ASO after preparing left ventricle or directly atrial switch procedure.[5]

While the postoperative management of these patients are usually straight forward, a small proportion of infants with transposition still experience a complicated and prolonged postoperative course following the ASO.[8] 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.[9]

In our country despite having huge number of patient burden, only one center is doing limited number of surgical management of TGA on a routine basis making it "Nouveau Espoir", the term comes from French language which means new hope. First successful outcome in ASO was also reported by Sharifuzzaman et al[10] from the same center in 2007. Other than these very few case reports, there are no literature published in our country relating with surgical management, post-operative issues as well as outcome of this disease in a resource limited set up. Given the scenario this study was aimed to show modalities of surgical treatment, document the immediate outcome, demonstrate major post-operative events and analyze our recent performance to track down improvement in outcome.

Materials and methods

This is a retrospective observational study, executed at National Heart Foundation Hospital & Research Institute (NHFH&RI) in the city of Dhaka, Bangladesh. The study sample consisted of all children as well as grown up and adult patients of both genders, diagnosed with TGA, subjected to cardiac surgery, during 2 years period from 1st July 2014 to 30th June 2016.

Data collection was executed from *"The Departmental Congenital Heart Surgery Database"* in the Department of Pediatric Cardiac Intensive Care. The data collection instrument approached issues related to: Demographic characters, diagnosis, procedures performed, cardiopulmonary bypass data, post-operative events, post-operative mechanical ventilation, ICU stay & post operative hospital stay, post operative inotrope score & vasoactive inotrope score and outcomes. *"The Departmental Congenital Heart Surgery Database"* of the Department of Pediatric Cardiac Intensive Care has been started since 1st July 2014. Every day one data entry operator entered the cases of the day and also the detail data entry of the patients who were shifted out of ICU. On the day of discharge pediatric intensivist again check all the data entry of that patient and complete the entry of that particular patient. To make the database having minimal error, periodic analysis of the database is also done.

During the period of time from 1st July 2014 to 30th June 2016 (2 years) at Pediatric Cardiac Intensive Care Unit we managed total 26 patients, who received surgical management for TGA. Congenitally Corrected TGA (ccTGA) patients were excluded from this study population. All data were computed using the Microsoft Excel 2010 software and analyzed with SPSS® statistical software version 16 (SPSS Inc., Chicago, IL). Continuous data are expressed as Median and range; categorical data are expressed as Percentage.

Results

There were total 26 patients in the study population, among them 2 patients were grown up and adult. The demographic characteristics of the rest of the studied patients (24 cases) showed that median age of the cohort was 2.1 months with a range of 0.9-16 months; median weight of 3.6 kg (range: 2.5-12 kg) and median length 100 cm (range: 45-83 cm) [Table-I].

Table-I

Demography (n=24)

	Median	Range
Age (months)	2.1	0.9 - 16
Weight (Kg)	3.6	2.5 - 12
Length (cm)	57	45 - 83

Table-II showed details of the two grown up and adult patients. First patient was a 10 years old male, a case of delayed presentation D-TGA, who was not suitable for anatomical correction, hence physiological correction (Senning procedure) was done. His post-operative period was uncomplicated and discharged home on 14th POD. Another patient was a case of delayed presented D-TGA with complex cardiac anatomy with extracardiac malformation unsuitable for single stage biventricular repair and finally palliative procedure was done (Modified BT shunt).

Table-II

Detail of two grown up and adult patients

Particulars	Diagnosis	Procedure	Post-operative events and outcome
10 years, Male	D-TGA, ASD secundum, anomalous origin of right coronary artery, pulmonary artery hypertension (PVRI 2.16 Wood)	Senning procedure	Uncomplicated post-operative period and discharged home on 14th post-operative day (POD)
24 years, Male	Dextrocardia, Situs inversus, D-TGA, Large secundum ASD L-R shunt, Large PM VSD with anterior and trabecular extension L-R shunt, Severe infundibular &valvular pulmonary stenosis with hypoplastic	Right sided B-T shunt (8 mm).	Heart failure due to shunt overflow, chylothorax treated with TPN and adult chylothorax diet, two episodes of culture positive sepsis (Acinetobacter and Enterobacter), re-exploration due to increased chest tube serous drainage, pressure sore over sacrum, Hypoal-buminemia, Generalized edema. Finally had full recovery and discharged home on 40th POD

PAs: Pulmonary arteries, L-R: Left to right, PVR: Pulmonary venous resistance, BT shunt: Blalock Taussig shunt.

Twenty one (81%) were male and 5(19%) female (Fig.-1).

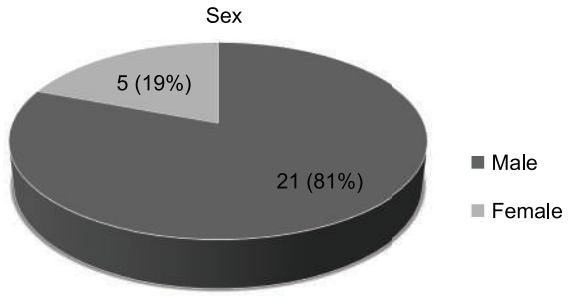

Fig.-1: *Sex distribution (n=26)*

Three patients (11.5%) required preoperative stabilization by emergency balloon atrial septostomy (Fig.-2).

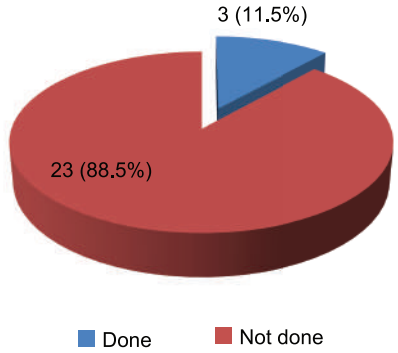

Fig.-2: *Preoperative procedure of Balloon Atrial Septostomy (BAS) [n=26]*

Half of the patients (50%) had TGA with intact ventricular septum (IVS), 9(34.6%) TGA with VSD, 3(11.5%) Taussig Bing anomaly and 1(3.8%) TGA with other complex cardiac anomaly (Fig.-3). Among all these patients ASO and ASO with VSD repair was done in 10(38.5%) patients each (Fig.-4). Other procedures were Senning procedure 4(15.4%), 1(3.8%) REV (Réparation à l' Etage Ventriculaire) procedure and 1(3.8%) palliative procedure (Modified BT shunt).

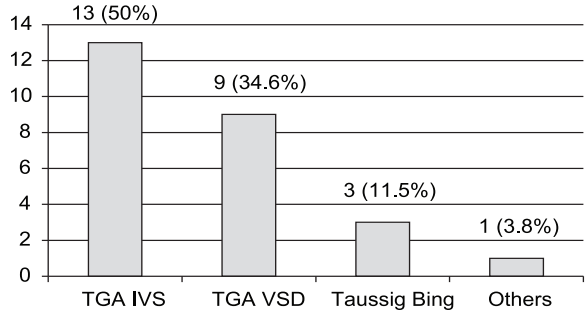

Fig.-3: *Types of TGA (Diagnosis) [n=26]*

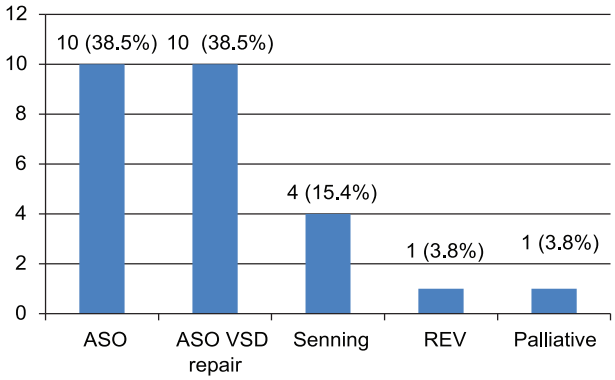

Fig.-4: *Procedures done (n=26)*

Among all the 26 cases, other than the Modified BT shunt, 25 surgeries were done under cardio-pulmonary bypass (CPB) with median CPB time 250 minutes (range 130-339 minutes) and median cross clamp time 149 minutes (range 94-205 minutes). Continuous ultra filtration (Cuff) was done in 24(92.3%) patients and modified ultra filtration was done in 23(88.5%) patients (Table-III).

Table-III
Perfusion data (n=25)

	Median (Range)	Number (%)
CPB time (min)	250(130 – 339)	
Cross clamp time (min)	149(94 – 205)	
Cuff		24(92.3)
Muff		23(88.5)

Nine (34.6%) patients came to ICU after surgery with open sternum and 6(23.1%) patients required re-exploration during post operative. All the patients required any form of inotrope infusion with median inotrop score (IS) of 7.25 (range 0-69.3) and median vasoactive inotrope score (VIS) 28 (range 5-84.7). Median duration of mechanical ventilation was 142 hours with a range of 16.2-742 hours. Ten (38.5%) patients required re-intubation. One (3.8%) patient developed pneumothorax which required tube thoracostomy and water seal drainage. Another 3(11.5%) patients required tube thoracostomy to manage significant pleural effusion. Three (11.5%) patients developed diaphragmatic palsy among them 2 improved spontaneously over time but 1 patient required diaphragmatic plication in view of repeated weaning failure from mechanical ventilation (Table-IV).

Table -IV

Major post operative events (n=26)

	Median (Range)	Number (%)	Comment
Open sternum		9(34.6)	
Re-exploration		6(23.1)	
Inotropes		26(100)	
Inotrope score (IS)	7.25 (0-69.3)		
Vasoactive IS	28 (5-84.7)		
Duration of MV (hours)	142 (16.2-742)		
Re-intubation		10(38.5)	
Pneumothorax		1(3.8)	
Pleural effusion		3(11.5)	
Diaphragmatic palsy		3(11.5)	Diaphragmatic Plication done in 1 (one) patient
C/S positive sesis		12(46.2)	
AKI		15(57.7)	3 patients require dialysis
Wound dehiscence		4(15.4)	

Most common cause of open sternum was increased bleeding in 5(19.2%) cases. Other causes of open sternum include bleeding plus LCOS, bleeding plus edematous heart, post cardiac arrest and unstable hemodynamics 1(3.8%) patient each (Table-V). Six (23.1%) patients required re-exploration during post operative period due to bleeding, LCOS, sudden cardiac arrest, profuse serous chest tube drainage, cardiac tamponade and mediastinitis in 1(3.8%) patient each (Table-VI).

Table-V

Causes of open sternum (n=9)

Causes of open sternum	Number %	Comment
Increased bleeding	5(19.2)	
Bleeding + LCOS*	1(3.8)	
Bleeding + Edematous heart	1(3.8)	
Post cardiac arrest	1(3.8)	Expired
Unstable hemodynamics	1(3.8)	Expired

***LCOS:** Low cardiac output syndrome

Table-VI

Causes of re-exploration (n=6)

Causes of re-exploration	Number %	Comment
Bleeding	1(3.8)	
LCOS*	1(3.8)	
Sudden cardiac arrest	1(3.8)	Expired
Huge chest tube drainage) (serous)	1(3.8)	
Cardiac tamponade	1(3.8)	
Mediastinitis	1(3.8)	

***LCOS:** Low cardiac output syndrome

One patient who underwent Senning procedure had complicated and stormy post operative course with bilateral temporoparietal ischemia, Junctional arrhythmia requiring anti-arrhythmic drugs, mediastinitis and prolonged mechanical ventilation requiring tracheostomy as well as prolonged post operative ICU stay. Another patient of Taussig Bing anomaly who underwent arterial switch operation had repeated weaning failure from mechanical ventilation due to pulmonary hemorrhage and finally improved after coil embolisation of 3 MAPCAs (Table-VII).

Table-VII

Other major post-operative events

Name of Procedure	Other major post operative events
Senning procedure	Bilateral temporoparietal ischemia, Junctional arrhythmia, Tracheostomy, mediastinitis requiring re-exploration, prolonged post operative ICU stay (106 days)
Taussig Bing Anomaly	Post operative MAPCA coiling (3 coils)

MAPCA: Major aorto-pulmonary collateral

Median duration of ICU was 12 days with a range from 1-106 days. We had 6 (23.1%) mortality among those patients (Table-VIII). Major causes of mortality include infection and LCOS. One patient died due to massive myocardial ischemia as a result of coronary abnormality. One patient after Senning procedure left hospital after surgery with residual neurological sequel as a result of bilateral temporo-parietal ischemia (Table-IX).

Table-VIII

Outcome (n=26)

	Median (Range)	Number (%)	Comment
Post operative ICU stay	12(1-106)		
Mortality		6(23.1)	
Discharged with morbidity		1(3.8)	One patient (Senning procedure) with residual neurological sequel

Table-IX

Details of the patient expired (n=6)

Serial	Age (m)	Diagnosis	Procedure	Causes of Death
1	2.2	TGA IVS	ASO	Severe LCOS
2	2.3	TGA VSD PS	REV procedure	LCOS, Septicemia
3	2	TGA VSD	ASO + VSD repair	Septic shock (Citrobacter Sp.) with DIC with AKI
4	3.3	Taussig Bing anomaly		ASO + VSD repair LCOS with Septicemia with DIC with AKI
5	1	TGA VSD	ASO + VSD repair	Septic shock, Peritonitis, Mediastinitis, AKI, sternal wound infection
6	2	TGA IVS	ASO	Massive myocardial ischemia

ASO: Arterial switch operation, IVS: Intact ventricular septum, PS: Pulmonary stenosis, DIC: Disseminated intravascular coagulation, LCOS: Low cardiac output syndrome, AKI: Acute kidney injury.

Discussion

The atrial switch operations (Senning procedure or Mustard procedure) were the first definitive repair for patients with TGA and produced good results.[11] Today the atrial switch is largely replaced by the arterial switch operation (ASO) with excellent outcome.[11,12] Although atrial switch is rarely performed today in the most of the advanced centers, it is not merely of historical interest as there remain a few important indications for this operation.[11] Moreover in many part of the world, in countries with emerging economics, patients present lately due to various reasons[7] leaving atrial switch as only option in TGA patients with regressed left ventricle. The median age of our patients reflected the same where patients presented at median age of 2.1 months and only one patient presented during neonatal period at 26 days. It is already proved that delayed diagnosis of congenital heart disease worsens preoperative condition and surgical outcome.[13,14]

TGA patients present with various anatomical forms with additional anatomical variations.[1,2] In our series we found TGA with IVS as most common variety (50%) followed by TGA VSD 34.6%, Taussig Bing anomaly 11.5% and 3.8% other variations. In a big series of 1200 patients of TGA, Losay et al[15] found TGA IVS, TGA VSD and Taussig Bing anomaly in 68.8%, 24.8% and 6.6% respectively. Other authors also showed similar findings.[9,16]

TGA Patients present early in neonatal period if there is inadequate mixing between pulmonary and systemic circulation.[1,2] Most hypoxemic neonates with D-TGA get benefit from early institution of prostaglandin E1 (PGE) alone. However, neonates with inadequate intracardiac mixing due to a restrictive foramen ovale will not improve solely on PGE. In this setting, the markedly increased pulmonary blood flow from the PDA may lead to deleterious left atrial hypertension, pulmonary congestion and low cardiac output requiring urgent BAS.[1,2] As pre-natal prediction of interatrial communication adequacy is imperfect, it is recommended that neonates with D-TGA be delivered in a center equipped to perform BAS.[5] In our study 11.5% patients had preoperative hemodynamic instability requiring BAS.

After surgery, 9 (34.6%) patients had open sternum in ICU for variable period of time. Reported literatures showed incidences of delayed sternal closure after ASO is one-quarter to one-third of patients[5], which is similar to our findings. Among our cases none of the open sternum was elective and in 1 of them sternum was kept open after an episode of hemodynamic deterioration and cardiac arrest in ICU. In a retrospective study, elective delayed sternal closure afforded no benefit in terms of mortality or morbidity.[17]

Cardio-pulmonary bypass (CPB) time is an important factor related with post operative outcome. According to Hanna et al[18], CPB time >240 minutes is associated with increased risk of death. CPB time (Median 250 minutes, range: 130-339 minutes) of the patients in this cohort is little higher which is probably due to relatively few number of cases operated per year in a resource limited setup. Literature also supports that CPB time and cross clamp time is lower in high volume centers.[19] On the other hand ASO for Taussig Bing anomaly needs longer CBP time due to additional anomalies.[20]

Significant number of patients in this study encountered some form of adverse post operative events which include re-exploration, re-intubation, pneumothorax, pleural effusion, diaphragmatic palsy, culture positive sepsis, acute kidney injury and wound dehiscence. Despite significant improvement in operative outcome, according to the Society of Thoracic Surgeons Congenital Heart Surgery Database, 40.6% of patients had ≥1 complication[21] after surgery for CHD. Major post-operative complications as high as 49.8-56.8% has been reported also in literature after ASO operation.[19]

Post operative period of ASO in simple TGA is mostly predictable.[8] Dibardino et al[9] showed in his series of ASO, median duration of mechanical ventilation was 3 days (range, 1 to 12 days) and median duration of inotropic support 4 days (range, 2 to 20 days), median ICU length of stay (LOS) 5 days (range, 2 to 38

days) and median hospital LOS 9 days (range, 3 to 42 days). On the other hand TGA with VSD and Taussig Bing anomaly may have little stormy post operative period due to pulmonary hypertension and additional cardiac abnormalities like arch anomaly.[20,22] In our series median duration of mechanical ventilation (MV) was 142 hours (range 16.2-742 hours), median post operative ICU stay was 12 days but few patients had prolonged stay which probably caused by high incidence of infectious complications (C/S positive sepsis in 46.2% cases). On the other hand delayed presented cases and mixed group of surgeries other than ASO are there contributing to prolonged period of MV and LOS. The maximum post operative stay was 106 days in a patient of Senning procedure who had stormy and eventful post operative period with ischemic stroke, mediastinitis, arrhythmia and prolonged mechanical ventilation. Factors independently associated with a prolonged postoperative stay in the cardiac intensive care unit after ASO operation include prematurity, difficulty feeding, capillary leak, need for preoperative inotropic support and postoperative infectious complications.[6,8] The use of an intraatrial baffling procedure such as the Mustard or Senning repairs for TGA has been associated with a high incidence of cardiac rhythm abnormalities.[12]

Surgical management of various forms of TGA showed promising outcome during recent years[5,6] where early outcome of ASO reached to mortality as low as 1%[18] at some high volume centers. Outcome of other surgeries for different anatomical variations of TGA also improving with 7% mortality in Arterial Switch Operation for Taussig Bing anomaly.[18,22] Although the early postoperative mortality 6 out of 26 patients (23.1%) in this cohort was higher than the recently reported series, there is significant improvement in comparison to our previous outcome of last 4 years from 2011-2014 where we had 17 mortalities out of 38 cases of ASO and ASO with VSD repair.[23] This improved outcome is probably related to a 'learning curve', employment of new techniques, improvement of perioperative care, CPB management and better post operative care, which is reflecting the same truth presented by Karamlou et al[19] showing center and surgeon volume each influencing outcomes after ASO.

Causes of death in this study are mostly due to LCOS and infectious complications. Reported post-operative complications causing worse outcome include unexplained profound ventricular dysfunction, low cardiac output syndrome or hemodynamically significant arrhythmias, including supraventricular tachycardia, junctional ectopic tachycardia or ventricular tachycardia, residual coronary abnormalities, myocardial ischemia, sepsis and pulmonary hypertensive crises.[12,18] The prevalence of coronary events in literature varies from 2% to 11%. The coronary events most often occur immediately after the ASO and are the main cause of death or morbidity. In the early postoperative period, coronary events are related to coronary anatomy and to surgical technique difficulties.[12]

Conclusion

Being a new center in a resource limited set up in a country from emerging economics, challenges are many to perform surgery in complex congenital heart diseases. Although over time our outcome is improving, there will be new challenges in future including more number of complex cases. Additional challenges include late presentation and high numbers of infectious complications. Early detection of the cases and prenatal diagnosis can help to bring better outcome.

Limitations

This is a retrospective review of small number of patient showing immediate outcome only. Longer duration of study with larger population with intermediate and long term follow up is required for stronger evidences.

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Pattern of Congenital Heart Diseases among Childern in Dhaka Shishu (Children) Hospital

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Abstract

Background: *Congenital heart diseases are the most common congenital problem in children and it is one of the leading causes of mortality in the first year of life. Presentation can vary from asymptomatic incidental finding to severe cardiac decompensation and death. The purpose of this study is to highlight the pattern of congenital heart diseases in children referred for echocardiography from out patient department and admitted patient of Dhaka Shishu hospital.*

Methodology: *This was a retrospective descriptive study on all patients with the confirmed diagnosis of congenital heart disease referred for echocardiography over a period of six months from July 2016 to December 2016. Patients from day one of life till 18 years were included. Study was conducted in the pediatric cardiology department at Dhaka Shishu Hospital. Data was entered in SPSS version 21 and analysis was done by using SPSS program.*

Results: *Out of 1721 referred suspected cases, congenital heart defects were detected in 981 patients. There were 56.6% male and 43.4% female. Acyanotic congenital heart diseases were observed in 78.3% of cases and cyanotic congenital heart diseases were found in 21.7% of cases. Ventricular septal defect was detected in 21.7% of cases followed by atrial septal defect 21.4%, Patent ductus arteriosus 10.4%, and AV canal defect 4.9% were the most common acyanotic congenital heart lesions. Whereas Tetralogy of fallot followed by D-TGA, DORV, Pulmonary atresia, TAPVC and Tricuspid atresia were the commonest cyanotic congenital heart lesions.*

Conclusions: *Ventricular septal defect was the most common form of acyanotic congenital heart diseases and Tetralogy of fallot was the most common form of cyanotic congenital heart diseases. Respiratory distress and cyanosis was most common symptoms of Congenital heart disease. 2D-echo with Doppler examination forms the gold standard for diagnosis.*

Key Words: *Congenital heart diseases, ventricular septal defect, tetralogy of fallot.*

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Introduction

Congenital heart diseases (CHD) is one of the most common structural malformation and comprises upto 25% of all congenital anomalies. It is one of the leading causes of mortality during the first year of life[1], and it is the most common human birth defect worldwide[2], it is prevalence approaching 1% of live births [3]. The incidence is higher in stillborns (34%), spontaneous abortions (10-25%), and

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premature infants (about 2% excluding patent ductus arteriosus (PDA).[4]

The causes of most congenital heart defects are unknown. Most cases of congenital heart disease were thought to be multifactorial and result from a combination of genetic predisposition and environmental factors. A small percentage of congenital heart lesions are related to chromosomal abnormalities particularly trisomy 21, 13, 18 and Turner syndrome. Heart disease is found in more than 90% of patients with trisomy 18, 50% of patients with trisomy 21, and 40% of those with Turner syndrome.

The risk of recurrence of congenital heart disease increases if a first degree relative (parent or sibling) is affected.[4,5] Two to four percent of cases of CHD are associated with maternal conditions and teratogenic factors, including maternal diabetes mellitus, phenylketonuria, systemic lupus erythematosus, congenital rubella syndrome, maternal obesity and maternal ingestion of drugs (lithium, ethanol, warfarin, thalidomide, antimetabolites, vitamin A derivatives and anticonvulsant agents).[6,7] Sex differences in the occurrence of specific cardiac lesions have been identified. Transposition of the great arteries (TGA) and left-sided obstructive lesions are slightly more common in males (>65%), whereas ASD, VSD, PDA, and pulmonary stenosis (PS) are more common in females.[4] Congenital cardiac diseases can be divided according the presence or absence of cyanosis into two major groups.

Acyanotic congenital heart diseases can be classified according to the predominant physiologic load that place on the heart. The most common lesions are those that produce a volume over load, and the most common of these are left-to-right shunt lesions such as ventricular septal defect (VSD), atrial septal defect (ASD). The second major class of lesions causes an increase in pressure load, most commonly secondary to ventricular outflow obstruction (pulmonary or aortic valve stenosis) or narrowing of one of the great vessels (coarctation of the aorta).[8] Cyanotic congenital heart diseases can be divided according to pathophysiology: whether pulmonary blood flow is decreased (Tetralogy of Fallot (TOF), pulmonary atresia with an intact septum, tricuspid atresia, total anomalous pulmonary venous return with obstruction) or increased (transposition of the great vessels (TGA), single ventricle, truncus arteriosus, total anomalous pulmonary venous return without obstruction).[8] The diagnosis can be confirmed by echocardiography, ECG, CT scan or MRI of the chest, or cardiac catheterization.[8] The aim of the present study was to determine the pattern of congenital heart disease among patient attending at Dhaka Shishu Hospital Echo room, its age and sex distribution, mode of presentation.

Materials and Methods

This is a retrospective study conducted in echocardiography room from July, 2016 to December, 2016 at Dhaka Shishu Hospital, Dhaka, Bangladesh. All children with the confirmed diagnosis of congenital heart disease were included. CHD is defined as the structural heart disease or intrathoracic great vessels that is actually or potentially of functional significance present at the time of birth even if there was a delay in detection, as defined by Mitchell et al[4].

Nine hundred and eighty one children were studied. Age ranged from day1 till 18 years of age. Clinical data were reviewed. Consideration was given to total number of cases with CHD, age at diagnosis, sex distribution and type of CHD. Patients with acquired heart diseases as rheumatic heart or mitral valve prolapse were not included in this study.

Results

During the study period, 1721 patients were referred for echocardiography, among them 981 patients (57.1.0%) were diagnosed as having CHD. There were 555 males and 426 females with male: female ratio of 1.3:1. The mean age was 11.5 months and mean weight was 6.56 kg.

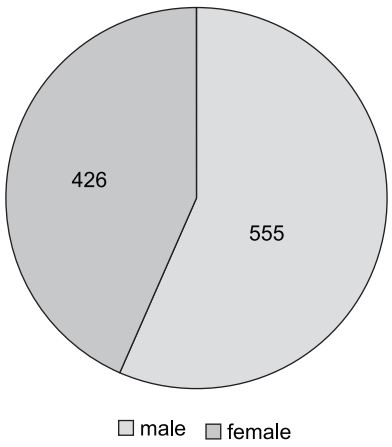

Fig.-1: *Sex ratio in congenital heart diseases (n=981)*

Seven hundred and sixty eight patients (78.3 %) had acyanotic CHD, while two hundred and thirteen patients (21.7%) had cyanotic type of CHD (Fig.-2).

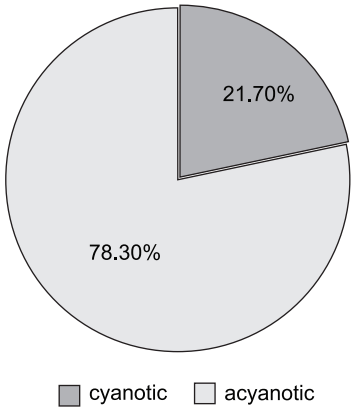

Fig.-2: *Type of congenital heart disease (n=981)*

In our study, isolated VSD was the most frequent form. It was found in 213 (21.7%) patients followed by ASD 210 (21.4%), PDA 102 (10.4%), AV Canal defect 48 (4.9%), VSD was associated with PDA in 36 patient (3.7%) and with ASD in 66 patients (6.7%). ASD associated with PDA 18 patient (1.8%). Among patients with obstructive lesion bicuspid aortic valve with aortic stenosis 18 patients (1.9%), pulmonary stenosis 9 patients (0.9%), coarctation of aorta 9 patients (0.9%). Among patients with cyanotic CHD, tetralogy of fallot (TOF) was the most frequent form of cyanotic CHD, occurring in 50 patients (5.2 % of the CHD) followed by D-TGA 40 patients (4.2%), DORV 24 patient(2.4%), Pulmonary atresia 21 (2.1%), TAPVC 18 patients (1.8%) Tricuspid atresia 15 patients (1.5%), Truncus arteriosus 12 patients (1.2%) [Table-I].

Table-I

Relative distribution of acyanotic and cyanotic CHD (n=981)

Cardiac lesion	Number	Percentage
Ventricular septal defect (VSD)	213	21.7%
Atrial septal defect (ASD)	210	21.4%
Patent ductus arteriosus (PDA)	102	10.4%
Atriventricular canal defect (AV canal defect)	48	4.9%
Coarctation of aorta	9	0.9%
Pulmonary stenosis (PS)	9	0.9%
Bicuspid aortic valve with Aortic stenosis (AS)	18	1.9%
Mitral regurgitation (MR)	27	2.8%
ASD+VSD	66	6.7%
ASD+PDA	18	1.8%
VSD+PDA	36	3.7%
VSD+PS	15	1.5%
Hypertrophic obstructive cardiomyopathy (HOCM)	6	0.6%
Corrected-Transposition of great artery	3	0.3%
Tetralogy of Fallot	50	5.2%
Transposition of great artery	40	4.2%
Double outlet right ventricle	24	2.4%
Pulmonary atresia	21	2.1%
Total anomalous pulmonary venous circulation	18	1.8%
Tricuspid atresia	15	1.5%
Truncus arteriosus	12	1.2%
Double inlet left ventricle	12	1.2%
TOF with absent pulmonary valve	6	0.6%
Ebstain anomaly	3	0.3%
Total	981	100%

It is evident that respiratory distress was the most common presentation of CHD which occurs in 399 patient (40.6%) followed by Cyanosis 117 patients (11.9%), recurrent chest infection with failure to thrive 102 patients (10.4%), respiratory distress with feeding difficulty 90 patients (9.2%), Incidental detection of murmur 51 patients (5.2%), shortness of breath on exertion 18 patient (1.8%) [Table-II].

Table-II

Symptoms in cases of congenital heart diseases (n=981)

Symptoms	No. of patients	% of Total
Respiratory distress	399	40.6
Cyanosis	117	11.9
Recurrent RTI + Failure to thrive	102	10.4
Feeding difficulty	84	8.6
Respiratory distress + feeding difficulty	90	9.2
Shortness of breath on exertion+ Cyanosis	18	1.8
Incidental murmur	51	5.2
Shortness of breath on exertion	27	2.8
Failure to thrive	54	5.5
Cyanotic spell	30	3.1
Cardiomegaly	6	0.6
Chest pain	3	0.3
Total	981	100%

Our study shows that 156 patients (15.9%) were neonates (<1 month), 615 patients (62.7%) were infants (1month - <12 months), 100 patients (10.2%) were toddlers (12 months- <36 months), 68 patients (6.9%) were in preschool age (36 months- <60 months) and 42 patients (4.3%) were in school age and above(5 years- <18 years) [Fig.-3].

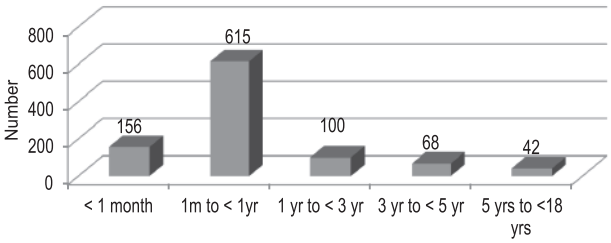

Fig.-3: *Age groups at the time of diagnosis of CHD*

Discussion

Congenital heart diseases are important group of diseases that causes great morbidity & mortality in children.[9] This study showed the relative frequency of various forms of CHD. There were 981 cases of CHD. Male to female ratio was 1.3:1.which is nearly similar to that reported from studies done in Dhaka CMH (1.39:1)[10], Bangabandhu Sheikh Mujib Medical University (BSMMU) (1.24:1)[10] and Jordan (1.28:1)[11] and slightly higher than that reported from Mosul City[12] which was 1.05:1, Saudi Arabia (0.9:1)[13] but Lower than reported from Dhaka Shishu Hospital (DSH) (1.98:1)[10] and National Institute of Cardiovascular Diseases (NICVD) (2.55:1)[10]. Seven hundred sixty eight patients (78.3%) were acyanotic and two hundred thirteen patients (21.7%) were cyanotic and this is almost similar to other studies done in Bangladesh[14] which was 78.5% acyanotic and 21.5% was cyanotic, Jordan[11] in which 74% of cases were acyanotic and 26% were cyanotic CHD and in Yemen[15] in which 85% of cases were acyanotic and 15% were cyanotic CHD. Another study done in Dhaka Combined Military Hospital (CMH)[10] and BSMMU[10], where acyanotic was 83.44%, 84.21% respectively and cyanotic was 16.56%, 15.79% respectively.

Ventricular septal defect was the most common form of CHD in the present study; it was seen in 21.7% of cases which was almost closer to study done in DSH[10,14] 25.7%, 26.9% and Dhaka CMH[10] (25.08%) but lower than other studies done in India 37%[16] Pakistan 29%[17] Nepal[18] 38%, in Nigeria[19] 55.1%, Iraq with different frequencies. It was seen in 54.8% of cases in Mosul[12], and in 52% of cases in Baghdad[20]. In other countries, VSD was still the most common form of CHD. It was seen in 33.9%

and 33.1% of cases in two different studies done in Saudi Arabia[9,13]. In Jordan[11] VSD was seen in 43.4% of cases, while in Turkey[21] VSD was seen in 32.6%, and 26.5% of cases observed in Yemen[15]. This difference in frequency of VSD in various studies could be attributed to differences in locality, time and study design.

In the present study the second most common form of CHD found was ASD. It was seen in 21.4 % of cases which was almost similar to study done in DSH 21%[10], 21.2%[14] and NICVD[10] (20.69%) but higher than that reported by other studies in India 12%[16], Pakistan 10.5%[17], Nepal 7.3%[18], Iraq 2.2% in a study done in Baghdad[20]. In other countries also ASD was seen in slightly lower frequencies than the present study. It was seen in 13.6% in a study in Jordan[11], 13.1% in a study in Turkey[21], and 12.2% of cases in Saudi Arabia[13].

In the present study PDA was seen in 10.4% of cases which was almost similar to study done in DSH[10] 10.66%, Jordan[11] 11% and Iraq 12.3%[12] but one study done in Nepal[18] seen lower frequency 7.5%. This is lower than that reported in other studies done in which was 14.9% in Pakistan[17], turkey (15.9%)[21] and in 13.6% in Baghdad[20], In Jordan[11], PDA was seen 8.3%, Saudia Arabia (6%) [13]. In the present study Atrioventricular canal defect was seen in 4.9% of cases which is similar to study done in DSH[14] 4.4%, Pakistan[17] 4.4% but lower in NICVD[10] 2.73%, BSMMU[10] 3.69% and Jordan[11] 3.6%. In the present study PS was seen in 0.9%%. This figure is lower than that reported from other studies done in Bangladesh[10,14], Iraq 5.3 % and 4.5 %[12,20], Jordan (6.2 %)[11], Saudi Arabia (12.4%)[9], Turkey (7.9%)[21]. The difference between figures of the present study and other study may be due to sample size used by the present study and other work.

In the present study coarctation of the aorta was seen in 0.9% of cases which was similar to one study done in Bangladesh[14] but lower than reported in other study in Nepal (1.8%)[18], Saudi Arabia (2.3%)[9], Pakistan (3.5%)[17], Jordan (3.4%)[11]. Tetralogy of Fallot (5.2%) was the most common form of cyanotic CHD in the present study, which is consistent to other studies done in BSMMU[10] 5.79%, Iraq[12,20], Jordan[11] and Saudi Arabia[22]. Second most common cyanotic congenital heart disease was d-TGA 4.7% which is similar to study done in Bangladesh[14] 4.8%, Jordan 5.5%[11] but lower than one study done in Pakistan 7%[17] and higher than one study done in Nepal 1.8%[18].

In the present study most of cases were diagnosed during infancy. This is similar to the studies done in Bangladesh[10,14], India[16], Pakistan[17], Nepal[18], Mosul[12], and Saudi Arabia[9].

Most common presentation was respiratory distrees (40.6%) followed by cyanosis 11.9%, FTT 10.4%, recurrent RTI 10.4% and incidental detection of murmur which is similar to one study done in Nepal[18], and another study done in Nigeria[19].

Conclusion

The majority of patients with congenital heart disease detected was acyanotic. VSD was the commonest acyanotic lesion and TOF was the commonest cyanotic lesion. Most of them are detected during infancy. The most common clinical presentation was respiratory distress. Early detection of CHD reduce the mortality and morbidity of patients.

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Review Article

Hypertension in Children: An Update

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Abstract

Prevalence of hypertension in children has increased significantly in recent times, in part related to the epidemic of childhood obesity. Identification and treatment of hypertension in childhood is likely to favorably impact on cardiovascular disease in adulthood. Identification of hypertensive children continues to be problematic because of incomplete blood pressure screening during routine pediatric clinical visits. The blood pressure norms are based on age, gender and height specific values in contradistinction to adults where a single value suffices. Childhood hypertension is either primary or secondary and is categorized as prehypertension (between 90th to 95th percentile), stage 1 (95th to 99th percentile plus 5 mmHg) and stage 2 (e" 99th percentile plus 5 mmHg) hypertension. Ambulatory blood pressure monitoring is useful in confirming the diagnosis and in helping diagnose white coat and masked hypertension. Once diagnosed as definitive hypertension, the causes of secondary hypertension should be determined and appropriately treated. In children with primary hypertension, a combination of life-style changes (diet and exercise) and drug therapy should be instituted depending upon the stage of the hypertension. Continued follow-up to ensure compliance with treatment regimen and to monitor blood pressure control is mandatory.

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Introduction

Prevalence of pediatric hypertension is increasing day by day. One possible explanation for this striking increase in prevalence over the past several decades is the concurrent rise in pediatric obesity. This significant increase makes it much more likely that clinicians will find themselves caring for hypertensive children, heightening the need for proper recognition, evaluation, and treatment in the primary care setting. Hypertension: It is defined as systolic blood pressure (SBP) and/or diastolic blood pressure (DBP) [3] 95th percentile for gender, age, and height on three or more separate occasions.[1] Normotensive is called when systolic blood pressure (SBP) and diastolic blood pressure (DBP) between 50th to 90th percentile for gender, age, and height.

Classification[1]

Prehypertension: Blood pressure (BP) levels that are in between 90th percentile to 95th percentile is termed prehypertension.

Stage 1 HTN: BP from the 95th to 99th percentile plus 5 mm Hg.

Stage 2 HTN: BP more than 99th percentile plus 5 mm Hg.

Hypertensive Urgency: Symptomatic elevated blood pressure without signs and symptoms of acute target organ abnormalities.

Hypertensive Emergency: Symptomatic elevated blood pressure with features of acute target organ abnormalities.

White coat hypertension (WCH): It is a condition where office or clinic BP is high but the BP during ambulatory blood pressure monitoring (ABPM) is normal.

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Masked HTN (MH): It is a high ABPM in the presence of normal office BP, is being recognized as a risk factor for cardiovascular complications in adults.

Dipping: {(mean wake BP -mean sleep BP)/mean wake BP}×100. Normal dipping is at least 10%.

Clinic BP index: clinic BP/95th centile for age sex height.

When to screen for hypertension:

- All children age 3 years and older should have their BP measured during each physician visit.
- Known risk factor where children <3 years old should have BP measured.

Techniques used to measure blood pressure in children are[1]

- Palpatory method
- Auscultatory
- Doppler Ultrasonogram
- Oscillomtery
- Ambulatory blood pressure measurement
- Direct, which requires an intra arterial catheter

How to measure blood pressure: The patient should be measured in basal state, child should be sitting comfortably quietly for at least 5 min. Measurement should be preferably in right arm in the cubital fossa at heart level.[1] Sometimes, particularly in a small or ill child, blood pressure is most conveniently measured in the leg with an arm cuff.[2] Selecting the right cuff size is also important. The inflatable bladder of the cuff, not the entire cuff, should be an appropriate size for the patient. The bladder width should be of approximately 40% of the arm circumference midway between the acromion and olecranon; the cuff should cover 80% of the circumference of the arm without overlapping; and the bladder width to length ratio should be at least 1:2.[3,4]

Following are the korotkoff phases of BP[5]:

Inflate cuff until palpable pulse disappears

K1 : corresponds to systolic BP

K2 : tapping disappears

K3 : tapping returns

K4 : tapping muffles

K5 : tapping disappears

In some young children no fifth sound can be heard, because sounds are still audible at 0 mm Hg. The new recommendation for these children is to use the fourth korotkoff sound (point at which the sound muffles) as the diastolic blood prssure.[6]

Ambulatory Blood Pressure(ABPM): ABPM is measured with a wearable oscillometric blood pressure device usually placed on the waist, which automatically measures and records blood pressure at prescribed frequent intervals (every 20 min when awake and every 30 min during sleep) over an entire 24-hours.[7]

Indications of ABPM measurement:

- Home BP in disagreement with clinic BP.
- Apparent drug resistant hypertension.
- Unusual BP variability.
- Determining the efficacy of drug treatment over 24 hours.
- Evaluating nocturnal hypertension.
- Chronic kidney disease, diabetes mellitus.
- Clinic BP index 1.0-1.2.

Advantages of ABPM: Allows multiple measurements typically during a 24 hour period and hence

provides a true picture of BP trends. It Identify white coat hypertension and masked hypertension. It also provides information regarding diurnal BP pattern and non dippers.[8]

Disadvantages of ABPM: It requires training and expertise in analysis and usually limited to children >5 years old due to lack of large series providing normative data.[9]

Risk factors of hypertension: Obesity, low socioeconomic condition, low birth weight, lack of breast feeding, sedentary life style and food habit like caffeine, salt intake, known renal and urologic diseases, recurrent urinary tract infections, systemic diseases like systemic lupus erythematosus, increase intracranial pressure, sleep apnea, smoking, history of neonatal intensive care admission, umbilical artery catheterization, turner syndrome, malignancy, history of transplant (kidney, bone marrow).[10]

Etiology of hypertension

Pediatric hypertension is categorized into two major types, primary (essential) and secondary hypertension. Of the causes of secondary hypertension vary with the age of the child. Overall, renal parenchymal or renal vascular causes of hypertension account for 70-90% of secondary causes.[3]

Etiology of hypertension

Renal causes[9,10]:
- Acute glomerulo-nephritis
- Chronic glomerulonephritis
- Acute kidney injury
- Chronic kidney disease
- Renovascular hypertension: renal artery stenosis, renal vein thrombosis, fibromuscular dysplasia.
- Vasculitis: systemic lupus erythematosus, takayassu arteritis, moyamoya disease.
- Hereditary nephropathy.
- Cystic kidney disease: autosomal recessive polycystic kidney (ARPKD), autosomal dominant polycystic kidney(ADPKD).
- wilms tumor.
- Reflux nephropathy.

Endocrine causes[9,10]:
- Long term corticosteroid intake: oral, parenteral
- Congenital adrenal hyperplasia
- Cushing syndrome
- Primary hyperaldosteronism (crohn's syndrome)
- Pheochromocytoma
- Hyperthyroidism

Cardiovascular causes[9,10]:
- Coarctation of aorta
- Arteriovenous fistula
- Patent ductus artreosus
- Middle aortic syndrome

Tumour[9,10]:
- Neuroblastoma
- Pheochro-mocytoma

Neurogenic[9,10]:
- Raised intracranial hypertension
- Gullain Barre Syndrome· Poliomyelitis
- Transverse

Medications[9,10]:
- Steroid
- Cyclospori-ner
- Non steroidal anti-inflamm-atory drugs
- Erythropoietin
- Cocaine
- Alcohol
- Albumin infusion
- Oral contraceptive
- Excessive

Clinical manifestation

Children is mostly asymptomatic. Children may have headache, neck pain, abdominal pain, nausea, vomiting, fatigue, epistaxis, blurred vision, Bell's palsy. Children can present with complications like left ventricular failure, encephalopathy, cerebrovascular accident, chronic kidney diseases, retinopathy. Left ventricular failure is presented by dyspnea, tachycardia, gallop rhythm, basal fine crackles. Encephalopathy is manifested by seizure, blurred vision, mental confusion, nausea, vomiting and headache.[3,9,10]

Neonatal presentation is different from older child. They can present with feeding difficulty, sepsis like features, dyspnea, congestive heart failure, irritability, lethargy, seizure and intracranial bleeding can be another mode of presentation. In the long standing case failure to thrive can be a feature.[9,10]

Primary hypertension

Essential (or primary) hypertension is often associated with a family history of hypertension or obesity. Family history of hypertension is present in nearly 80% of patients with primary hypertension and is thought to be due to multiple genetic and environmental factors. The prevalence of primary hypertension is higher in native american, hispanic, african american and lower in asian children.[5-7]

Clues for secondary causes (four extremity BPs or target organ damage, such as hypertensive retinal changes via fundoscopic examination) should be searched for. Obesity with BMI greater than the 95th percentile for age and gender is often associated with chronic primary hypertension.[11] The presence of peripheral edema, pleural effusion, rash, or swelling and tenderness of joints suggest acute onset or exacerbation of underlying renal or systemic disease. Retinal changes usually indicate long-standing, untreated hypertension and include arteriolar narrowing or tortuosity, arterio-venous nicking, hemorrhages and exudates· Metabolic syndrome mean combination of hypertension, obesity and dyslipidemia.[12]

Physical clues suggest underlying cause of hypertension[13]

Growth retardation, anemina and distress suggestive of chronic renal failure. Truncal obesity, moon facies, acne, hirsutism and striae suggest cushing syndrome. Abdomen mass, tachycardia suggest neuroblastoma. Palpable mass, tachycardia, flushing, diaphoresis and pallor may be due to pheochromocytoma. palpable kidney can be due to polycystic kidney disease, multicystic dysplastic kidney, hydronephrosis. Epigastric /flank bruit may be due to renal artery stenosis. Decreased lower extremities pulses, drop in BP from upper extremities suggest coarctation of the aorta, takayasu arteritis . Webbed neck, widely spaced nipples is features of turner syndrome. Thyromegaly suggest hyperthyroidism. Ambiguous /virilization may be due to adrenal hyperplasia. Caf-au-lait spots due to neurofibromatosis and Adenoma sebaceum may be due to tuberous sclerosis. Malar rash may suggest systemic lupus erythematosus. Adenotonsillar hypertrophy may cause sleep apnea, snoring.

Investigations

As renal disease is the most common cause of hypertension in children and the kidney is also a target organ for damage from untreated hypertension, urinalysis is an important screening test. Other test to rule out glomerulonephritis, quantification of protein by protein creatinine ratio or 24 hours total protein. Hematuria and red cell casts with or without proteinuria suggests glomerular disease. Isolated hematuria also may be associated with dilatation of the urinary tract or trauma. Proteinuria may be seen in non-glomerular conditions such as reflux nephropathy, obstructive uropathy, or interstitial nephritis, as well as glomerular diseases like focal segmental glomerulo- sclerosis or membranoproliferative glomerulonephritis. To find out the cause of vasculitis complement factor C3 and C4, antineutrophilic antibody (ANA), anti double strand antibodies(anti-ds DNA), perinuclar anineutrophilic cytoplasmic antibody. Other initial investigations should include blood urea, scrum creatinine, scrum electrolytes sodium, potassium, chloride, bicarbonate, calcium, phosphate, uric acid. Elevated serum potassium in conjunction with metabolic acidosis may suggest chronic renal disease, which is confirmed by the presence of an increased serum creatinine concentration and estimation of creatinine clearance.[14] The renal ultrasound provides information about the size and architecture of each kidney and the lower urinary tract. Abnormal kidneys may be small or asymmetric (renovascular disease, vesicoureteral reflux or dysplasia); hyperechoic, symmetric and normal or large (renal parenchymal disease like glomerulonephritis) or large with or without cysts (polycystic kidney disease, multicystic dysplastic kidney). Nuclear imaging dimrcaptosuccinic acid scan (DMSA), diethyl triamine penta acitic acid (DTPA), renal biopsy may be required. Hypokalemia concentration with metabolic alkalosis may indicate like primary or secondary hyperaldosteronism or Liddle syndrome. Duplex ultrasonography provides information about the patency and flow within the main renal vessels to help diagnose renovascular hypertension. Selective renal arteriography which is the gold standard for diagnosis of renal artery stenosis[3] Computed tomographic angiography (CTA) and magnetic resonance angiography (MRA) are 80-90% accurate alternatives to renal arteriogram for evaluation of renovascular disease. Plasma renin activity, plasma and urine catecholamines for patient with hypokalemia, previous umbilical catheter placement or other risk for renovascular hypertension.[15,16] For determination of target organ injury echocardiogram(ECG) should be done. For estimation of co morbidities fasting glucose and fasting lipids should be done.[16]

Management

It includes pharmacological, non pharmacological and rarely surgical means.

Goal of Therapy

1. keep the BP less than 95th percentile if there is no other coexisting disorder.
2. Keep the BP less than 90th percentile in children with evidence of coexisting cardiovascular abnormality.[17]

Pharmacological management

All children with HTN and diabetes should be considered for pharmacologic therapy, independent of albuminuria status.[15] Childhood hypertension is usually secondary, pharmacotherapy needs to be started. Following antihypertensive drugs are used frequently.

Oral dose of anti hypertensive drugs[3]

Drug	Dose	Maximum dose
Calcium channel antagonist		
Amlodipine	0.05mg/kg/day 24 H	0.2mg/kg/day
Nifdipine	0.25mg/kg/dose 6-8H	0.5mg/kg/day
Diltiazem	1.5–2 mg/kg/day 8 H	
Verapamil	3–8 mg/kg/day 8 H	
Felodipine	2.5 mg/day 6-8 H	
Isradipine		
ACE inhibitors		
Captopril	0.1 mg/kg/dose 8H	6ng/kg/day 8H
Enalapril	0.08 mg/kg/dose 12-24H	0.5mg/kg/dose 12H
Fosinopril		
Quinapril	5–10 mg/day	
Lisinopril	0.07 mg/kg per day upto 5 mg/day	
Ramipril	6mg/m^2/day	
Angiotensin ii receptor antagonist		
Losatan potassium	0.5 mg/kg/dose 24H	1.4 mg/kg/dose 24H
Irbesartan	2 mg/kg/dose	
Candesartan	0.23–0.35 mg/kg per day	
Valsartan	1.3 mg/kg/day (q.d.)	40 mg/day
Olmesartan	0.3 mg/kg per day	
Beta blocker		
Atenolol	0.5 mg/kg/dose 12-24H	8 mg/kg/dose 8H
Propanolol	0.2-0.5 mg/kg/dose 6-12H	2 mg/kg/dose 6-12H
Metoprolol	1–2 mg/kg/per day 12 H	
Bisoprolol	2.5/6.25 mg/day	
Alpha and beta blocker		
Labetalol	1-2 mg/kg/dose 12H	10mg/kg/dose 6H
Carvedilol	0.08 mg/kg/dose 12H	0.75 mg/kg/dose 12H
Diuretics		
Furosmide	0.5-1 mg/kg/dose 6-24H	12 mg/kg/day
Hydrochlorothiazide	1-1.5 mg/kg/dose 12-24H	4mg/kg/day
Metolazone		
Spiranolacton	: 1 mg/kg per day	
Triamterene	1-2mg/kg/day	
Amiloride		
Bumetanide	0.015–0.06 mg/kg/day 12 H	
Epleronone	0.5–1 mg/kg per day	
Peripheral Alpha agonist		
Prazosin	0.025-0.1 mg/kg/dose	0.5 mg/kg/dose 6-12H
Terazosin	1mg/day	
Central Alpha agonist		
Clonidine	5-10mcg/kg/day 8-12H	0.9mg/day
Vasodilators		
Hydralazine	Initial: 0.75 mg/kg/day 6-8 H	
Minoxidil		

There are three general guidelines

1. Further increase the dosage gradually to the maximum tolerable dose ("stepped" or "step-up" care), or,
2. Replace the initial agent with that of another class ("sequential monotherapy") on the assumption that it is not possible to predict how all children with a given disorder will respond to any individual agent, or,
3. Add another agent ("add-on" care).[16]

There are three essential steps to aid the decision to choose a specific class of antihypertensive medication and which agent within a class for a particular child:

Step 1. Formulate a hypothesis of the Pathophysiology of HTN and of contributing factors. This will aid the selection of one or more agents with a mechanism of action that may counteract or oppose such pathophysiologic mechanisms.

Step 2. Identify comorbid conditions such as diabetes and other metabolic disorders, heart disease, pulmonary or kidney disorders and consider whether or not these disorders may be negatively impacted by the proposed agents.

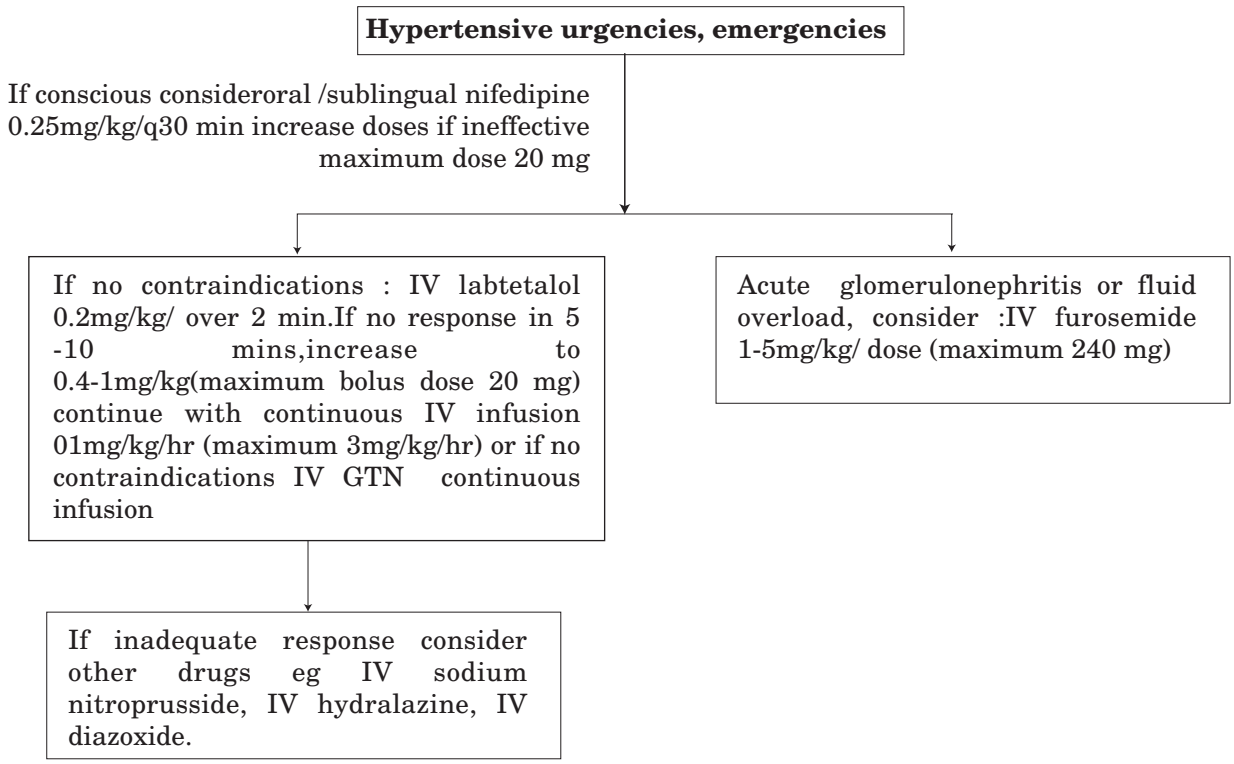
Step 3. Monitor for adverse effects associated with specific antihypertensive agents.[18]

Non-pharmacologic management of HTN

Lifestyle changes are essential in managing all children with HTN. The NHBPEP recommends a 1.2 g/day sodium intake for 4-8 year-olds and 1.5 g/day for older children[19] (figure 1). A daily aerobic activity lasting 30–60 min is highly recommended. Static exercises are less useful for weight control while excessive weightlifting (more than 30 min/day for more than 3 days/week) may contribute to HTN. Limit TV and other sedentary activities.[20]

Figure 1: *Healthy eating plate for hypertension*

Management of Hypertensive emergencies



In acute hypertension first 1/3 of total BP reduction to be aimed for over 8 hour, next 1/3 over over 12 hour and final 1/3 over 24 hours. Some of the intravenous antihypertensive medications used in the treatment of hypertensive emergencies include sodium nitroprusside, nicardipine, esmolol, hydralazine, labetalol, fenoldopam, and phentolamine. Some of the medications used in the treatment of hypertensive urgencies include enalapril, nifedipine, clonidine, minoxidil, and angiotensin II receptor blockers.[21]

Conclusions

HTN in children are commonly secondary and hence underlying cause should be searched and treated. Hypertension resolves when acute treatable secondary causes resolves. Primary HTN are increasing with along with obesity, they can be prevented by changing life style and food habit.

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Case Report

Two-Patch Repair for Atrioventricular Septal Defect without Down's Syndrome: A Case Report

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Abstract

We experienced an unusual case of complete atrioventricular septal defect in a 13 month old patient. A preoperative echocardiogram revealed complete balanced AV canal defect Rastelli A type, large ostium primum ASD, moderate inlet VSD with upper muscular extension and left to right shunt, severe PH, moderate to severe right and left AV valve regurgitation. A crescent-shaped PTFE patch was placed beneath the inlet VSD and a section of autologus pericardium was place in the primum ASD.

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Introduction

There are numerous variations in anatomy of the atrioventricular connection in atrioventricular septal defects (AVSDs).[1,2] 2D and Doppler echocardiogram is the most common and useful examination to evaluate such structures[3], however, it does not always provide fine detail or allow for accurate diagnosis of pathology. Atrio-ventricular septal defects (AVSD) comprise a spectrum of malformations which were recently classified by the Congenital Heart Surgeons Society. Characteristic pathological findings in this entity of malformations of the endocardial cushion tissue are an ostium primum ASD, a cleft in the anterior leaflet of the mitral valve, and in complete AVSD, an inlet VSD with a common atrioventricular valve (Fig.1). The two septal defects are only separated by the common AV-valve, which varies with regard to the anatomy of the superior bridging leaflets as classified by Rastelli into types A, B, and C. AVSD malformations are common cardiac defects; a complete AVSD is the typical cardiac malformation in patients with trisomy 21 and up to two thirds of patients with this malformation have trisomy 21.[4] We present a 13 month old case of complete Rastelli Type A AVSD without Trisomy 21 in this report.

Case Report

A 13-month-old female child was referred to our department from Pediatric Cardiology Unit for surgical repair of a complete AV canal

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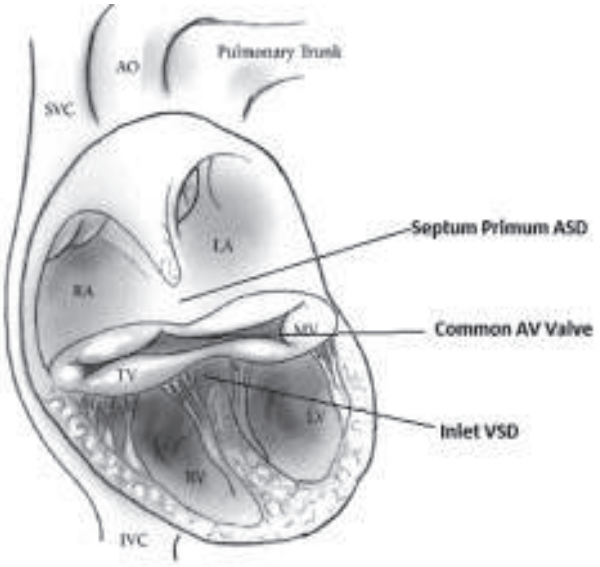


Fig.-1: *Complete AV canal defect*

defect Rastelli type A. The 2D and Doppler echocardiogram showed a septum primum ASD and a deficient inlet type of ventricular septum beneath the atrioventricular valve (Fig.-2). This anomaly was diagnosed as complete AVSD, however, clinically the child did not have any feature of Down's Syndrome which was also confirmed by doing karyo-type, AV canal defect is a congenital cardiac defect commonly associated with Down's Syndrome, opposite to that in this case it is a rare association without Trisomy 21. There was

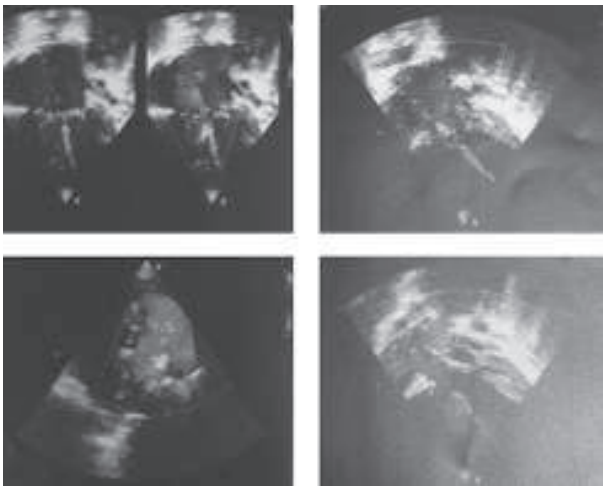


Fig.-2: *Doppler Eco-cardiogram of AVSD*

also moderate to severe right and left valve regurgitation was seen. Systolic pressure in the pulmonary artery was 40 mm Hg (approximately 43% of systemic pressure), and the ratio of pulmonary artery flow to systemic flow was 2.6. Chest X ray shows pulmonary plethora and increased transverse diameter of the heart (Fig.3). We also performed a cardiac catheterization with oxymetry which reveals a large VSD and normal coronaries. All pulmonary veins drained into the left atrium.

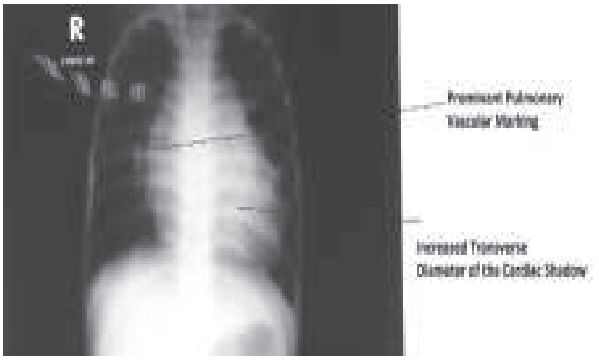


Fig.-3: *Chest X-ray P/A view of AVSD*

No PLSVC (Persistant left superior vena cava). Pulmonary vascular resistance before giving 100% oxygen was 3.57 wood Units/M^2 and after giving 100% oxygen it was came down to 2.96 wood Units/M^2.

We decided to go for operation and two-patch repair, which is normally performed for complete AVSD, was appropriate for this pathology.

During the operation, the pathologic anatomy was investigated by way of a right atriotomy under ordinary cardiopulmonary bypass and cardioplegic arrest. The ventricular septum under the atrioventricular valve was deficient, and there was a common AV valve and a septum primum ASD found over the common valve as was described in the Eco-cardiogram. The defect was Complete AV canal defect of Rastelli type A because there was a VSD and Primum type ASD, common AV valve, and slight bridging of left inferior leaflet of left AV valve (Fig.4).

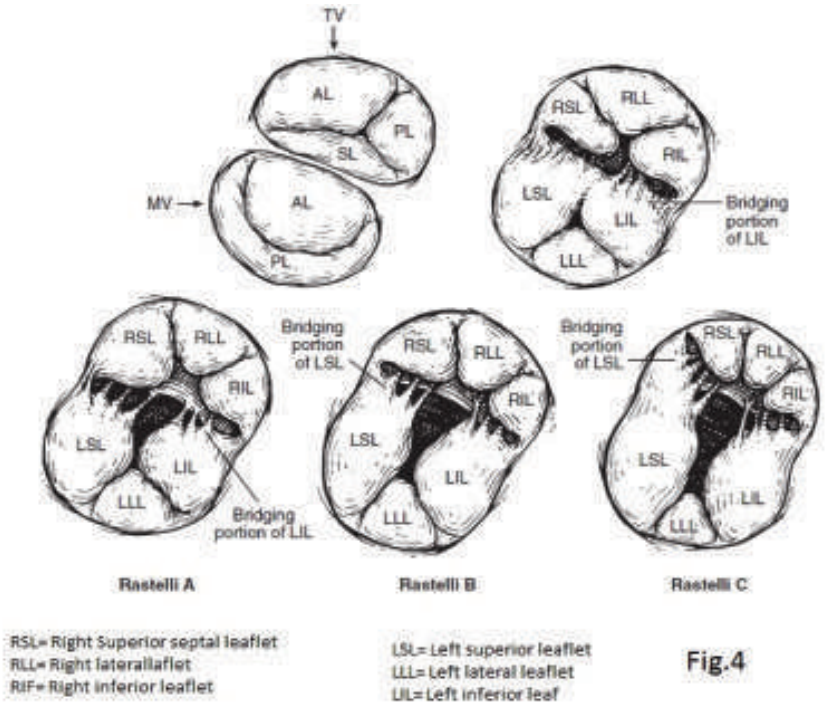
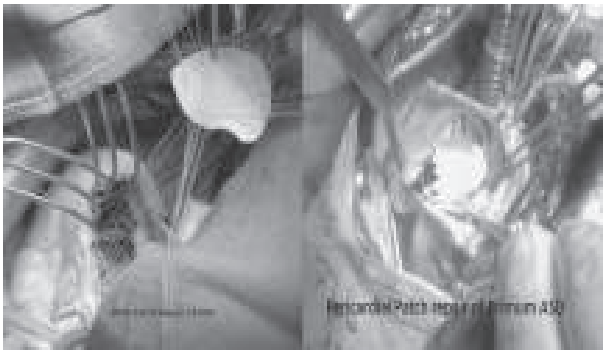


Fig.4

First, mid sternotomy was done, both cavae (SVC & IVC) and, aorta were cannulated directly with thin-walled, right-angled metal cannula and a straight cannula, aortic root cardioplegia cannula also inserted. CPB flow was reduced to about 1.2 L/min/m^2 when the patient's temperature reduced up to 30°C. As cooling proceeds, the aorta was clamped, and cold cardioplegic solution was injected. The right atrium was opened widely, and a sump sucker was passed through the foramen ovale into the left atrium. Stay sutures were applied. The malformation was examined and each morphologic detail noted. Morphology of the LSL and LIL was noted carefully with the leaflets in both the closed and open positions. Cold saline solution was injected through the valve and the closure pattern and any regurgitant leaks observed. The most anterior point of LSL-LIL opposing edges was found, and a double-armed 7-0 polypropylene suture was placed through it. Leaflet stay and marking sutures were placed, measurements were made, and the PTFE interventricular patch was trimmed. The patch was sutured to the right side of the crest of the ventricular septum with continuous polypropylene suture. Chordae of the RSL and RIL stayed on the right ventricular side of the patch; those of the LSL and LIL stayed on the LV side, and some were cut when they interfered with the suturing, because the anterior edges of these leaflets would be sutured to the PTFE patch. When this phase was completed, the marking suture on the anterior edges of the coapting surfaces of the LSL-LIL complex was passed through the appropriate point of the edge of the PTFE ventricular defect patch. The pericardial interatrial patch was then trimmed to appropriate shape and size, and the first part of its insertion was accomplished. For this, interrupted mattress sutures of 6-0 proline were placed to enclose anterior edges of the LSL and LIL between the PTFE patch below and pericardial patch above.



Great care was taken to ensure alignment of left AV valve leaflets was perfect and without distortion during this process. Saline solution was again injected through the left-sided portion

of the AV valves (two orifices were present once the interventricular patch was in place) to study its closure pattern and competence, minimum regurgitation and well coaptation of valves were noticed. The diameter of each was estimated with Hegar dilators and considered acceptable Repair was completed by suturing the rest of the pericardial interatrial patch in place, with the suture line passing around the AV node and bundle of His and not across them. A few sutures were placed for closure of the foramen ovale. Air was evacuated through the foramen ovale, and the aortic clamp was removed with suction on the needle vent. Atriotomy was closed and the patient weaned from Cardio-pulmonary bypass without any adverse event.

Discussion

We believe that this is the first report presenting in our institution as a AVSD without Down's Syndrome, most of which were previously reported, had an association with Down's Syndrome[5,6] occurring in AVSD patients. In the present case, a child without Down's syndrome presented with failure to thrive and features of cardiac failure, echocardiography revealed complete balanced AV canal defect and cardiac Cath report was favorable for operative repair. Our preoperative diagnosis was almost accurate and decision to operate on this patient was prompt and judicious without any delay to avoid further complications. In fact, a previous report contained similar Eco-cardiographic observation, although a different diagnosis was reached.[7]

A deficient cordal structure and leaflet malformation are always present in the atrioventricular valve of AVSD.[8] Distinguishing mitral and tricuspid tissues in the atrioventricular valve was difficult. However, once the left ventricle was volume loaded, only the mitral portion of the atrioventricular valve was bulged; thereafter, the mitral and tricuspid areas were easily identified and the bridging portion of the mitral leaflet could be recognized. To determine the size of the PTFE patch and its attachment line on the mitral leaflet, the anterior mitral leaflet had to be bulged.

Although this technique is not normally used for partial AVSD, we attached a PTFE patch underneath the atrioventricular valve to secure the closure of the shunt from the left ventricle, normalize the height of the atrioventricular valve, and obtain adequate mitral coaptation by pushing back the mitral leaflet to the left side. Because left atrioventricular valve regurgitation is an unresolved problem[9], creation of a new mitral annulus has certain benefits for further mitral surgery. The prosthetic mitral ring can be easily placed in future if ever necessary, on this new annulus without injuring the conduction system.

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Right arch dominant of double aortic arch with Tetralogy of Fallot

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Abstract

A 4 years old male child presented to us with a history of repeated cyanotic spell and episodes of squatting for last 6 months. There was no history of compression symptoms in the form of dysphagia, recurrent regurgitation, and cough. A routine chest X-ray showed a right ventricular apex and pulmonary oligemia. Echocardiography revealed TOF, with right sided aortic arch with suspected double aortic arch (DAA). CT angiogram revealed a double aortic arch with TOF with normal coronary pattern. The patient underwent total correction with separation of double aortic arch.

Key words: *Tetralogy of Fallot, Double aortic arch.*

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Introduction

Double aortic arch is the most common form of complete vascular ring, encircling both the trachea and esophagus, resulting in non-cardiac morbidity like stridor, dyspnea, cough, recurrent respiratory tract infection and dysphagia.[1] Asymptomatic DAA with tetralogy of Fallot is extremely rare.[2] We present a case report of this rare anomaly associated with TOF which was treated successfully by separation of DAA followed by total correction of TOF by a midline sternotomy only.

Case report

A 4 years old male child presented to us with a history of repeated cyanotic spell, episodes of squatting for last 6 months and increasing cyanosis. There was no history of compression symptoms in the form of dysphagia, recurrent regurgitation, and cough. Physical examination revealed deep cyanosis (SPO_2 80%), grade III clubbing and an ejection systolic murmur (grade 4/6) at pulmonary area. A routine chest X-ray showed right ventricular apex and pulmonary oligemia with normal viscerobronchial situs with gastric air bubble on the left side and a liver shadow on right side. In echocardiography, the patient had situs solitus and levocardia; there was a large subaortic ventricular septal defect (VSD) with aortic override. There was severe infundibular & valvular PS (PPG 74 mm of Hg). It also revealed suspected DAA. DAA was confirmed on computed tomography angiogram. It revealed DAA with TOF; right arch 7.6 mm containing right common carotid and subclavian artery and left arch 9.9 mm congaing left common carotid and left subclavian artery. There was no compression sign on trachea also (Fig. 1). Mc Goon ratio, Nakata index and mitral and tricuspid valve ratio was 1.8, 213 mm^2/BSA and 0.63 consecutively. With these investigations, the patient was taken up for surgery.

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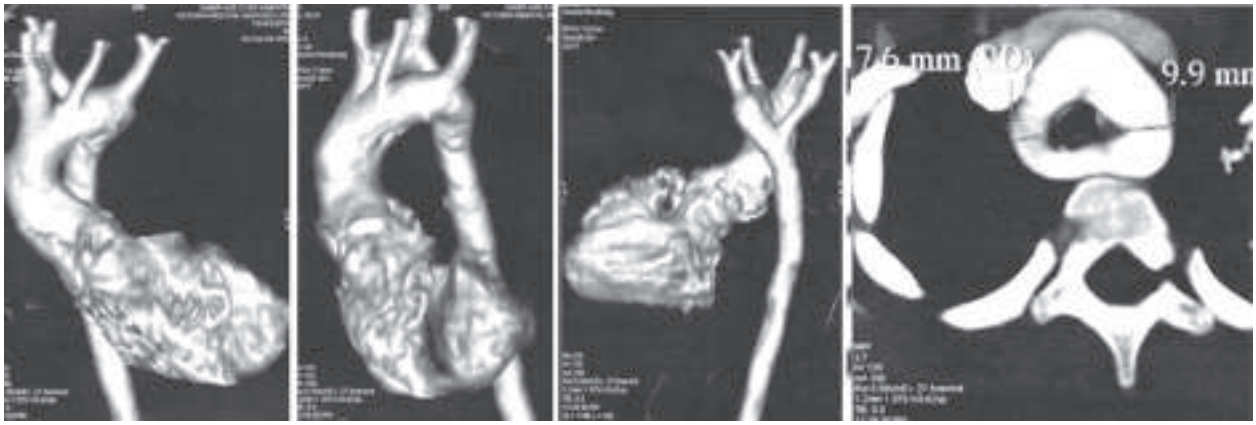

Fig. 1: *CT angiogram of Heart & great vessels revealed DAA.*

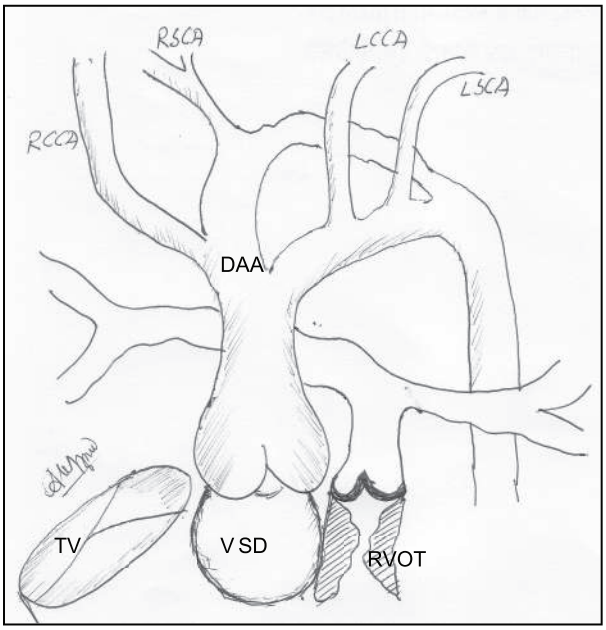

Fig. 2: *Peroperative morphology*

After induction, three arterial lines were given (right & left radial and femoral). Intraoperatively, a midline sternotomy was done. Anatomy was checked peroperatively (Fig. 2). Extrapericardial aorta was dissected free up to the thoracic spine, behind the trachea and oesophagous. Total correction of TOF (VSD repair with Dacron patch, RVOT muscle band resection, MPA augmentation with pericardial patch) was done under standard Cardio-pulmonary bypass (CPB) and cooled to 26^0C. To detect the dominant arch, we clamped the both anterior (right) and posterior (left) arch to find out femoral arterial pressure changes. After clamping the posterior arch (left) distal to right subclavian artery, there was no pressure change between left radial and femoral artery. So, anterior arch (right) was dominant for descending thoracic aorta (DTA). On low flow and TCA, posterior arch (left) between right subclavian and DTA was clamped and divided after taking purse string at each end and ligation of both end was done with 6/0 prolene (Fig. 3a & 3b). Postoperative recovery was uneventful.

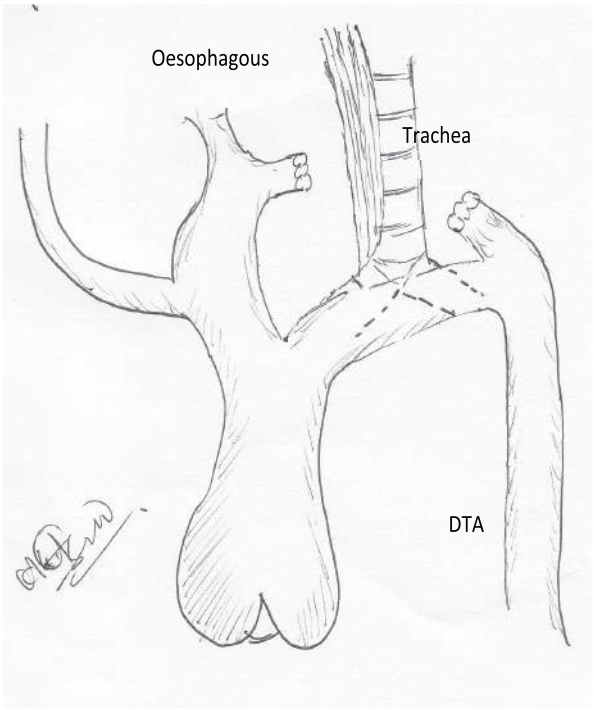

Fig. 3b: *After correction (CT angiogram)*

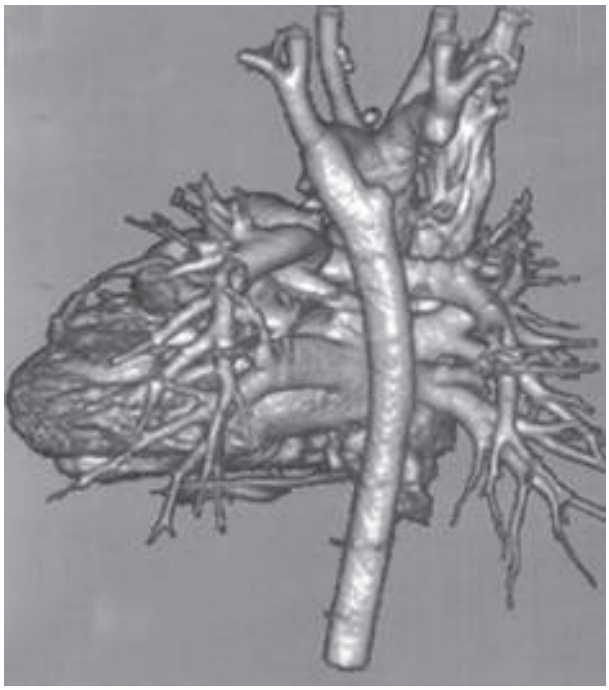

Fig. 3a: *After correction*

Discussion

Double aortic arch can be classically separated into three groups: dominant right aortic arch, dominant left aortic arch, and balanced aortic arch. Most of the patients have the dominant right arch type of DAA. The incidence rate of patients who have dominant right and left arches is only approximately 5%.[3-5] Right arch dominant DAA was detected in our patient.

Echocardiogram is useful to evaluate the aortic arch and heart anatomy. It is noninvasive, readily available and allows accurate delineation and exclusion of other major cardiac pathologies. Supra-sternal echo-cardiography view most of the times provides accurate information.[6] Currently, CT and magnetic resonance (MR) imaging showed more accuracy for the evaluation of mediastinal structures.[7] They provide excellent preoperative definition with no discrepancies with surgical findings.[6] In our case, CT angiogram provides excellent information regarding DAA and its surgical management.

Patient in the present case did not have any symptoms due to DAA but was probably symptomatic due to TOF with severe infundibular and valvular pulmonary stenosis. Surgical repair in the present case was performed after taking consent from the patient and posterior arch was ligated. His subsequent course in ICU was satisfactory. But, on 8th postoperative day he developed generalized tonic clonic seizure which was managed with phenobarbitone. All postoperative biochemical markers and CT scan of brain was normal and there was no further episode of seizure. During discharge, he was fully conscious with normal behavioral state and no focal deficit.

Conclusion

Asymptomatic right arch dominant type of DAA with tetralogy of Fallot is extremely rare. High index of clinical suspicion supplemented with stepwise radiological approach can clinch the diagnosis in most cases. This prevents the early and long-term respiratory and gastrointestinal complications. Outcomes are excellent after repair of DAA in most centers.

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