Diagnosis and Initial Management of Heart Disease in the Newborn

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INTRODUCTION

The incidence of congenital heart disease (CHD) is estimated at 0.4 to 0.8 % in studies from various parts of the world.[1-7] Our large population of over one billion clearly underscores the magnitude of the problem of CHD. Many congenital heart defects can now undergo definitive surgical or transcatheter procedures with excellent immediate and long-term results. This is particularly true if we intervene early. For these reasons, the focus of paediatric cardiologists has shifted to the infant and newborn, and many congenital cardiac lesions are now addressed in this age group. The facilities comprehensive neonatal cardiac care, are, in particular, are very much limited in India as of now but the situation is likely to change with the establishment of a number of pediatric cardiac centers.[8,9] The total numbers newborns in India with heart disease that receive treatment probably represent a tiny fraction of the total number of newborns with critical CHD (CHD that is usually fatal without early specific intervention) in the country.[7] Thus, the vast majority newborns with serious CHD are probably dying before they could be referred to a centre with expertise.

Heart diseases that manifest during newborn period very often require urgent attention. Prompt treatment can often yield gratifying results and many instances excellent long-term event free survival can be expected. Today, some form of palliative or definitive treatment is feasible for most newborns with CHD. Table 25.1 lists the common cardiac emergencies at birth.

Successful management of a newborn with congenital heart disease requires careful attention to a number of steps starting with recognition of heart disease in the newborn to initial resuscitation, transport to a pediatric cardiac center, assessment and detailed diagnosis and palliative or definitive procedure at the pediatric cardiac center. This chapter outlines the principles involved in each of the steps.

Table 25.1: Cardiac emergencies in the newborn

Physiologic category	*Conditions*	*Manifestation*
Duct dependent systemic blood flow* failure, acidosis	Hypoplastic left heart syndrome, critical coarctation, interruption of aortic arch, critical aortic stenosis	Heart failure, shock, circulatory
Duct dependent pulmonary blood flow*	Pulmonary atresia, critical pulmonary stenosis, Ebstein anomaly	Cyanosis, hypoxia
Obstruction of pulmonary venous return	Obstructed total anomalous pulmonary venous return, mitral atresia with a restrictive patent foramen ovale	Cyanosis, hypoxia, heart failure
Parallel circulation with poor mixing	D transposition with intact ventricular septum	Cyanosis, hypoxia
Valve regurgitation	Congenital mitral valve regurgitation	Heart failure
High-output state	AV malformations (usually intracranial)	Heart failure
Myocardial dysfunction	Myocardial diseases (inflammatory and metabolic)	Heart Failure

Tachyarrhythmia	Atrial flutter, neonatal atrioventricular reentrant tachycardias, ectopic trial tachycardia	Tachycardia, heart failure
Bradyarrhythmia	Complete heart block	Bradycardia, heart failure

*Some of the duct dependent conditions (critical PS, AS, coarctation) manifest with varying severity. The most severe forms manifest early (in the first few days) with absolute dependence on the duct for survival. Others may manifest later in the neonatal period (first few weeks) with heart failure or cyanosis and may not be strictly "duct dependent".

DIAGNOSIS OF HEART DISEASE IN THE NEWBORN

One of the major reasons for the delay in referral of infants with significant heart disease is failure to suspect heart disease in the newborn baby at initial clinical evaluation. Clinical diagnosis of heart disease in the newborn can be quite challenging. Manifestations of potentially life threatening CHD are often subtle and can be confused with noncardiac conditions. For instance, low cardiac output states resulting from critical aortic stenosis or other left-sided obstructive lesions may be mistaken for sepsis. Unfortunately the cost of failure to recognise CHD is, not infrequently, death. This is because many forms of CHD that manifest in the early neonatal period are fatal without specific interventions. It is possible to recognize CHD through careful clinical evaluation using the principles outlined below and a few additional tests. The diagnostic strategy for suspected CHD in the newborn is largely dictated by the condition of the newborn.

Clinical Clues

Clinical clues are often subtle. However, the trained pediatrician or neonatologist often picks them consistently. Persistent tachypnea (respiratory rate > 60 /min) is a consistent finding in most serious congenital heart defects in newborns. Over a few hours of observation it is often possible distinguish transient from persistent tachypnea. Evidence of respiratory distress in the form of grunting respiration, intercostal or subcostal retractions, flaring of alae nasae do not consistently accompany tachypnea. Cyanosis of extremities and lips is sometimes not easy to identify. Apart from a trained eye, there is the need for good lighting. Any suspicion of cyanosis should be verified by measurement of oxygen saturation.

Cardiovascular Examination

A thorough and systematic cardiovascular examination provides valuable clues to the presence of heart disease. With practice such an examination can be accomplished in a short time. For the pediatrician, a thorough familiarization with what is normal is a useful initial step. It is useful to answer the following questions that can serve as a checklist for a preliminary cardiac examination. This checklist is not at all comprehensive and is designed primarily for answering the question: Does the patient have heart disease? It can also help, identify the broad physiologic category of heart defect.

Are the Arterial Pulses Normal?

- Is the pulse volume normal or increased?
- Is there a discrepancy of pulsation in any of the four extremities?

A careful evaluation of pulses in all extremities should always be a part of physical examination. Coarctation is readily diagnosed when weak femoral pulses are detected in comparison to brachial or radial pulses. While a four extremity blood pressure measurement should ideally accompany clinical evaluation in all children with suspected heart disease, it is not always feasible. Nonetheless, when a discrepancy in pulses is suspected, four extremity blood pressure measurements should be obtained. An automated noninvasive blood pressure instrument is preferred over manual recording for four extremity blood pressure measurements.

Does the Precordium Feel Normal?

- Are there visible precordial pulses?
- Is the apex beat displaced, hyperdynamic or heaving?
- Is there a thrill palpable?

Are the Heart Sounds Normal?

- Can the two components of the second heart sound be separated?
- Is there an additional heart sound in diastole (such as S3)?
- Are there any additional systolic sounds (ejection clicks)?

Is (or are) there a Murmur (or murmurs)? If so:

- Is it systolic or diastolic or both systolic and diastolic (diastolic murmurs always have a structural basis)?
- Is it loud (grade 3 or louder murmurs are seldom innocent)?

Oxygen Saturation

In view of relative lack of sensitivity of the routine clinical examination for detection of CHD and the potential implications is not diagnosing critical CHD at birth, recent studies have focused on the use of pulse oxymetry for detection of CHD. Koppel et al reported a sensitivity of 60% and specificity of 99.95%, for pulse oxymetry in detecting significant CHD.[10] Similar results have been reported in other Studies as well.[11, 12] Most of these studies have used of cutoff of < 95% saturation as indicative of significant CHD. Hoke et al reported that lower limb saturation less than 92% in room air or 7% lower than that in the upper extremity could suggest critical left heart obstructive disease in the newborn.[13]

It is important to pay attention to details while recording oxygen saturations. There should be good correlation between the heart rate recorded in the pulse oximeter and the actual heart rates in the ECG monitor. The measured oxygen saturation has to remain consistent (variations of less than 1 to 3%) over 30 seconds

The ECG and Chest X-ray

Beyond the neonatal period, a normal ECG and chest X-ray makes the diagnosis of a hemodynamically significant heart defect unlikely. In newborns, however, ECG and chest X-ray changes, may take a few days to evolve. For the newborn, particularly in the first few days a "normal" ECG and a normal X-ray *does not rule* out serious heart disease. Notwithstanding this important caveat, important clues can often be obtained from the chest X-ray and ECG.

Like in any situation the chest X-ray needs to be looked at systematically. Specific attention needs to be paid to heart size, lung vascularity and, the situs. Although certain conditions are associated with specific cardiac contours (e.g. the "egg on side" contour in transposition), their specificity is very low. The presence of a right aortic arch is a useful clue and may be seen in patients with tetralogy of Fallot's or persistent truncus arteriosus.

In young infants, assessing heart disease is often difficult because of a large thymus. The thymus is characteristically a soft structure in the anterior mediastinum, which imprints the anterior cartilaginous portion of the ribs and creates a characteristic undulating or rippled appearance on the anteroposterior film. Occasionally, a lateral film is necessary to ensure that the structure of the thymus is in the anterior mediastinum and that indeed the heart is not enlarged. On a lateral chest radiograph of a normal child, a line from the trachea to the diaphragm does not

usually intersect the heart. On the other hand, when cardiomegaly is present, the heart is posterior to the line.

The radiographic assessment of pulmonary blood flow is difficult even for the experienced observer but it is even more difficult in an infant where the arteries and veins are small. Furthermore, pulmonary arterial flow in the neonate may be misleading because the patient may have a patent ductus arteriosus. Pulmonary venous hypertension typically occurs in association with obstructed TAPVC. This conditions often manifests on the chest X-ray as a ground-glass haze that is often mistaken for hyaline membrane disease.

The Hyperoxia Test

The hyperoxia test is frequently used to rule out critical CHD (conditions that are fatal without specific interventions in the newborn period). Examples of conditions that may be picked up by an abnormal hyperoxia test include transposition of great arteries, obstructed total anomalous pulmonary venous connection (TAPVC), conditions that are dependent on the patent ductus arteriosus for survival such as critical pulmonary stenosis, pulmonary atresia with or without a VSD, interrupted aortic arch, severe coarctation, hypoplastic left heart syndrome and critical aortic stenosis. This test is based on the principle that administration of 100% oxygen can raise the PO_2 of the arterial blood to a much higher level in absence of shunting from cardiac causes. It requires estimation of PO_2. This can be obtained from an arterial blood gas sample or, if available, a transcutaneous PO_2 probe. A cut-off of 300 mm Hg after 10 minutes of 100% oxygen administered via a hood (or the endotracheal tube in inubated patients) has been recommended. Values of 300 mm Hg or above virtually rule out CHD. A PO_2 of 200 mm Hg or more is also unusual for most forms of CHD, although a few conditions like persistent truncus arteriosus may have marked increase in pulmonary blood flow that can overcome the reduction in PO_2 from the admixture physiology. Values below 100 mm Hg strongly suggest congenital heart disease. In conditions like transposition, pulmonary atresia and obstructed TAPVC, PO_2 seldom rises above 70 mm Hg. During a hyperoxia test, it is important to include a sample from a "post-ductal" site. Either femoral artery or the umbilical artery can be used for arterial blood gas samples. For a transcutaneous probe the lower quadrant of the abdomen may be used. Absence of adequate standardization is the single biggest limitation of the hyperoxia test. Not uncommonly, PO_2 values between 100 and 200 mm Hg are obtained. This constitutes a grey zone where the results need to be viewed in context of other clinical findings.

The Echocardiogram

When doubts persist whether a patient has CHD or not despite a thorough clinical exam and chest X-ray, ECG, and hyperoxia test, an echocardiogram should be arranged. The threshold for obtaining an echocardiogram is much lower in institutions that have ready access to the investigation. This is especially true if the patient's condition does not permit adequate clinical evaluation. Such a situation is not infrequent in neonates and in such emergencies, it may be appropriate to bypass obtaining an ECG and chest X-ray. Portable and hand held echocardiography is now feasible and is increasingly being used. It is also now possible to train the paediatricians and neonatologists to perform a basic echocardiography screen for serious congenital heart disease.

Initial resuscitation and stabilization of a newborn with suspected heart disease.

Airway and Respiratory Support

Like in any other emergency situation a stable airway needs to be established first. Newborns with severe respiratory distress should immediately receive bag and mask ventilation and 100% oxygen may be used for this purpose (although, later the oxygen concentration may need to be reduced).

If respiratory distress continues to be profound after the initial resuscitation the newborn should be intubated and mechanical ventilation should be initiated. Neuromuscular blockade, sedation and atropine are recommended prior to intubation. Even in the most emergent situations sufficient time is often available to organize the requirements for performing intubation. It is useful to memorize the following checklist of items required for performing endotracheal intubation expeditiously:

1. *Control of the airway and preoxygenation with bag and Mask:* Make sure that the Ambu bag and mask of appropriate size is available. Test the Ambu bag and make sure that it works well, the bag does not leak and the valve works well. If the physician is alone at the time of intubation, she/he should first take control of the airway and give instructions to ensure that points 2 to 9 listed below are all carried out.
2. *Suction:* Choose a suction catheter of appropriate size. Make sure that the suction apparatus works.
3. *Access:* Ensure that a reliable peripheral or central venous access has been obtained.
4. *Monitoring:* ECG monitoring and monitoring of oxygen saturation throughout the procedure by pulse oxymeter is required and this should be instituted prior to endotracheal intubation.
5. *Medications:* All medications required for the intubation should be available. It is strongly recommended in newborns with heart disease that intubation should be performed after sedation and neuromuscular paralysis to avoid unnecessary stress on the cardiovascular system. It is a good idea to standardize a few protocols for drugs and use them for most cases. All dosages should be planned in advance and the drugs should be loaded and kept ready. Medications include atropine (0.02 mg/kg), ketamine:1 to 2 mg/kg bolus dose, (alternatives: Morphine: 0.1 to 0.2 mg/kg IV with or without Midazolam: 0.1 to 0.2 mg /Kg), neuromuscular paralytic agents: (Vecuronium 0.1 to 0.2 mg/kg or succinylcholine 2 mg/kg)
6. *The Laryngoscope:* Straight and curved blades of various sizes.
7. *The endotracheal tube and stilette:* For newborns, endotracheal tubes (sizes 2.5, 3 and 3.5 mm) should be available and kept ready. Typically for a term newborn size 3 works in most situations. Occasionally the airways will safely allow the use of a 3.5 mm tube. The 2.5 mm tube may be required for small preterm newborns.
8. Maggil forceps if elective nasotracheal intubation is planned or if an orotracheal position is to be changed to a nasotracheal position.
9. Items required for fixation of the tube.

The decision to intubate the newborn and initiate mechanical ventilation electively prior to transport requires consideration of the following variables: condition of the newborn in terms of severity of cyanosis, hemodynamic stability, gestational age and transport distance.

Access

A secure peripheral or central intravenous access is very essential. Inotropic agents with vasoconstrictor properties like dopamine and adrenaline can only be administered via a reliable central access. Extravasation of these agents into the subcutaneous tissue can result in extensive tissue necrosis in the event a peripheral line leaks. In the newborn infant, for the first 3 to 5 days, an umbilical venous line can be easily obtained. Alternatives include a jugular or a subclavian access. Generally both these routes are somewhat difficult and require some expertise and experience, particularly in centers without an infant or a neonatal surgical program. The femoral route should be avoided at all costs before the cardiac diagnosis is established. This is because the

either femoral vein may be required for cardiac catheterization in the immediate or distant future. For the same reason the femoral artery also should not be cannulated and repeated blood sampling should not be attempted from either groin. If central access is unavailable and an inotrope needs to be infused, dobutamine may be a reasonable choice.

It may be impractical to obtain arterial access prior to transport and unnecessary arterial punctures for blood gas sampling should be avoided because these sites will be required for placement of an arterial line prior to definitive surgery. An arterial sample is however necessary for ABG analysis. If the institution where the child is initially resuscitated has a blood gas analysis facility, an umbilical arterial sample should be obtained.

Oxygen

The potential dangers of excessive oxygen in a newborn with suspected heart disease include acceleration of closure of the ductus arteriosus and unacceptable decline in the pulmonary vascular resistance. Both these situations can have catastrophic consequences. Duct closure is fatal in duct dependent lesions. A marked decline in pulmonary vascular resistance translates into excessive pulmonary blood flow, often at the cost of reduced systemic blood flow. This is particularly likely to happen in duct dependent conditions. For these reasons, the FiO_2 needs to be titrated to maintain an oxygen saturation of 85 ± 5%. In most situations this allows a reasonable balance between pulmonary and systemic blood flows.

Prostaglandin — Availability, Administration and Alternatives

Prostaglandin E1 (available in India as Prostin VR) is a very essential drug and should be available in every newborn nursery. It can restore ductal patency in most newborns with closing ducts and is therefore life saving in duct dependent situations. Its effect is usually confirmed by improving saturations in newborns with duct dependent pulmonary circulation and resolution of the circulatory failure and acidosis in newborns with duct dependent systemic circulation. Its efficacy declines somewhat with increasing age particularly after 15 days and it is usually not effective in opening a closed duct after 30 days. The initial dose of prostaglandin is 0.05 to 0.1 microgram/kg/minute. Once the duct has opened up (this can be confirmed by the clinical response or by echocardiography), the dose may be reduced to as low as 0.01 mcg/kg/min. This allows maintenance of ductal patency with minimal adverse effects. Adverse effects of PGE1 infusion include apnea, bradycardia, tachycardia, hypotension, fever, gastric distension and seizures. Leukocytosis frequently accompanies prostaglandin use. Administration over several days may result in increased lung and body water from capillary leak, thrombocytopenia, gastric outlet obstruction and cortical hyperosteosis.

Prostaglandin is available in most cities in India. Newborn nurseries should endeavor to obtain one or two ampoules of the drug and this should be stored in the refrigerator and replaced when consumed. The drug is expensive (~ Rs. 6000 to 8000). It is, however, possible to extend the use of a single ampoule to about a week by using only small amounts of the drug to prepare the infusion on a daily basis. The remainder of the drug can be aspirated from the ampoule under strict sterile precautions (preferably under a laminar flow system) and stored in a sealed 1-cc syringe for as long as a week. Reducing the maintenance dose to a minimum can also help prolong the availability.

In the absence of prostaglandin, atropine 0.02 mg/kg boluses may be used as an alternative. For patients with transposition and intact ventricular septum, an umbilical venous catheter may be passed into the left atrium under fluoroscopic guidance and this could “stent” the atrial septum open to maintain reasonable oxygen saturations until transport to a center for balloon septostomy can be accomplished.

CIRCULATORY SUPPORT AND INOTROPES

Colloid or Crystalloids

Hypotension after PGE1 infusion is common. It is the result of relative intravascular volume depletion because of a combination of peripheral vasodilation and increased vascular permeability. It is best treated by administration of 10 to 20 ml/kg of a colloid solution (5% albumin or plasma) as a bolus. If colloid solutions are unavailable, crystalloid solutions may be used in the same volume. Inotropes are likely to be ineffective unless adequate volume replacement is done.

Dopamine

This should only be administered via a central line. Doses range from 5 to 15 mcg/kg. This is often the first choice in most centers. It has vasoconstrictor as well as inotropic effects and is often the only agent necessary.

Dobutamine

Perhaps the only reason to use dobutamine during initial resuscitation is that it done not require a central access. It has a potent inotropic effect and some vasodilatory effects. For this reason it is not as effective as dopamine in hypotension, particularly if the myocardial contractility is normal.

Adrenaline

It has powerful vasoconstrictor and inotropic effects but is seldom required as an infusion prior to or during transportation unless the neonate sustains severe hypotension or a cardiac arrest. Central access is a must and doses range from 0.01 to 0.2 mcg/kg/min.

Isoproterenol

Newborns with congenital complete heart block can be born with severe bradycardia and infusions of isoproterenol (0.01 to 0.05 mcg/kg/min) may be initiated prior to transport to center with facilities for pacemaker implantation.

Basic Laboratory Tests

Conditions that compromise systemic circulation (such as hypoplastic left heart syndrome) are likely to have an impact on liver and renal functions. A baseline ABG is very useful. Impaired systemic circulation is likely to result in metabolic acidosis. Conditions such as transposition and duct dependent pulmonary circulation and obstructed TAPVC are like to be associated with significant hypoxia. A septic screen (CBC, CRP, micro-ESR and blood cultures) should be performed at a low threshold. Sepsis in a newborn can have a significant impact on the immediate management strategies after referral.

TRANSPORTATION OF SICK NEWBORN INFANTS WITH HEART DISEASE

Communication

The decision to transport a newborn to a tertiary referral center with facilities for specialized care of neonates and infants with heart disease should be a joint one involving the referring pediatrician and the pediatric cardiology team. A thorough communication of important information that is obtained at the referring center helps to reduce problems during transport and helps to prepare the pediatric cardiac center to receive the newborn. Emergency procedures can

be planned on arrival with minimal delay. For example, if a newborn is being transported with a diagnosis of transposition, the cath lab can be prepared to perform a balloon septostomy with minimal delay after arrival. The following list could serve as a checklist of points to be communicated by a pediatrician or a neonatologist to the pediatric cardiology center prior to transportation of a newborn with suspected heart disease.

Prenatal Background

Term or preterm, birth order, age of mother, important maternal conditions.

Birth

Mode of delivery, relevant antenatal issues, Apgar scores, birth weight, significant postnatal events if any?

Clinical presentation: Why was heart disease suspected?

Current condition: Vitals (heart rate, respiratory rate, oxygen saturation, peripheral circulation), lab tests (a baseline arterial blood gas analysis report is valuable, if available), feeding.

Preliminary Diagnosis of the Cardiovascular Condition

Results of the physical examination, chest X-ray, ECG, and echocardiogram.

What has been done so far to resuscitate the newborn? What access has been obtained? Is the child ventilated or breathing spontaneously? What medications have been given (specifically, inotropes, prostaglandin)?

Relevant socioeconomic issues: What is the social and economic background of the family? Which family members are likely to accompany the child? How much has been communicated to them?

Logistics: Transport distance, mode of transport and transporting personnel? Expected time of departure and arrival?

Personnel for Neonatal Transport

Whenever feasible, the newborn should be accompanied by the resident neonatologist or pediatrician taking care of the baby and a nurse. Both the team members should be familiar with the underlying condition and should be aware of the potential problems the newborn may face during transport.

Monitoring during Transport

Ideally, it is necessary to continuously monitor ECG and oxygen saturations during transportation. This may not always be feasible, particularly in our environment. Often it is only realistic to monitor vital signs. Keeping a regular watch on respiration, heart rates and the overall general condition may be all that is feasible. When the newborn reaches the referral center, it is necessary to summarize the condition during transport and indicate important events, if any that occurred during transport.

Care of the Newborn during Transport

A secure airway and an access are vital. Newborns on prostaglandin can have periods of apnea. For this reason a number of units in the west would routinely intubate and mechanically ventilate a newborn on prostaglandin infusion during transport. For a number of reasons this is

not practical in the Indian scenario. Transportation while on prostaglandin infusion does amount to taking a calculated risk. The physician involved with the transport should be alert to this possibility and be ready to support respiration with a bag and a mask.

Excessive oxygen (> 2l/min/ FiO_2 > 40%) should be avoided during transport of most newborns with heart disease. This is probably advisable even if PGE1 is used to keep the duct open. High FiO_2 reduces pulmonary vascular resistance and can result in excessive pulmonary blood flow at the cost of systemic circulation. This is particularly dangerous in hypoplastic left heart syndrome.

Attention to other basic details such as temperature control, fluid balance, avoidance of hypoglycemia and hypocalcemia are all mandatory as in any neonatal transport situations.

Asepsis

Sepsis frequently complicates the management of newborns with heart disease. The potential for nosocomial sepsis is particularly high for sick newborns that are being transported. A number of caregivers are likely to handle the child and invasion in the form of endotracheal intubation, central and peripheral line insertion are likely to have taken place. Meticulous attention to aseptic precautions is extremely important. All nurses and physicians handling the newborn have to be specifically instructed as many of them may not be routinely used to newborn transport.

Evaluation and Management at the Tertiary Care Center

Detailed Diagnostic Evaluation

After receiving the newborn and ensuring hemodynamic and respiratory stability, a comprehensive echocardiogram must be performed. This should not be a rushed study. No matter what the initial diagnosis is, the cardiologist performing the echo must pay attention to all components of a comprehensive checklist of items. These include: visceral and atrial situs, systemic and pulmonary veins, the atria, atrial septum, ventricles, ventricular septum, conotruncus, great vessels, aortic arch, and the ductus arteriosus. Once the heart defect(s) is/are identified, the physiology needs to be thoroughly described as well. Patients with tachyarrhythmia in the newborn period should undergo a detailed evaluation that should clearly identify the basic mechanism of the arrhythmia. This allows appropriate treatment (Table 25.2).

Table 25.2: Specific treatment for neonatal cardiac emergencies

Conditions	*Specific Treatment*	*Comment*
Hypoplastic left heart syndrome mortality, often not because of significant and requirement of	Stage I palliation (Norwood operation)	Significant surgical practical in India long-term problems multiple operations
Critical coarctation/interruption over balloon dilation (lower restenosis rates) outcomes	Primary surgical repair of aortic arch	Surgery is preferred in coarctation excellent long-term
Critical aortic stenosis –term results. Can 90 to 95% success rate	Balloon valvotomy	Excellent intermediate be carried out with
Pulmonary atresia–intact precise anatomic ventricular septum hypoplasia and dimension,	BT-Shunt, radiofrequency wire puncture of atretic pulmonary valve followed by balloon dilation or surgical restoration by RVOT patch, stenting of the	Treatment dictated by issues (extent of RV tricuspid valve annulus

ventricle dependent circulation) and experience of the	patent arterial duct may also be required in addition.	presence of right coronary center
Pulmonary atresia with VSD or later in life dictated tetralogy of Fallot	Emergency BT shunt or stenting of the arterial duct other defects, "severe"	Definitive procedure by precise anatomy
Critical pulmonary stenosis outcomes. 5 to 10% mortality	Balloon valvotomy	Excellent long-term immediate
Obstructed total anomalous outcome; immediate 10 to 30%	Emergency surgical repair pulmonary venous return	Good long-term surgical mortality of
D transposition with intact to be performed in ventricular septum after which a Senning perhaps advisable	Balloon septostomy followed by arterial switch operation	Arterial switch needs the first 21 to 28 days, operation is
Vein of Galen AV fistula, embolization are generally AV malformations (usually are multiple intracranial)	Transcatheter coil, embolization	Results with coil poor especially if there communications
Myocardial diseases (inflammatory ACE inhibitors, improve with time	Medical management and metabolic)	Digoxin, diuretics and many patients
Tachyarrhythmias precise arrhythmia	Appropriate antiarrhythmic medications identification of underlying mechanism of	Treatment dictated by the
Complete heart block implantation is the first few days of life and outcomes can be	Temporary pacing followed by permanent pacemaker	Permanent pacemaker implantation feasible in excellent long-term expected

Assessment of End-organ Insult

Preliminary investigations to identify the extent of end-organ injury must be carried out. These include liver and renal function tests. Typically, conditions presenting with shock or severe hypoxia are associated with varying degrees of liver enzyme and renal parameter elevation. Ideally these patients are stabilized over the next several days to allow for recovery before definitive treatment is undertaken.

Ruling out Sepsis

It is vital to rule out ongoing sepsis whenever a newborn with suspected heart disease is received in a tertiary care center. A sepsis screen in the form of ESR, C reactive protein estimation, complete blood counts should be performed for all patients irrespective of their condition on arrival. It is reasonable to obtain a blood culture in all such newborns. Prophylactic antibiotics are generally not indicated unless the child shows clinical signs of sepsis.

Specific Treatment at the Tertiary Center

Specific treatment options in the tertiary center are largely dictated by the precise diagnosis and are listed in Table 25.2.

CONCLUSION

It is reasonable to expect a good outcome for most newborns with heart disease after the initial definitive or palliative procedure. Timely detection of heart disease after birth and appropriate referral is the vital initial step. Careful attention to small details during the initial resuscitation and transport is also vital particularly if the newborn is acutely ill. At the referral institution, a detailed and thorough assessment of the heart disease should be followed by institution of the appropriate specific treatment strategy.

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