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The Neonate with Congenital Heart Disease : Diagnosis and Management

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In the neonate cyanotic lesions as well as acyanotic lesions (left to right shunts and critical obstructions) manifest in several ways and identification of such lesions is critical in this age group. Evaluation of various presenting signs and symptoms along with various investigational modalities available which includes oxygen saturation, echocardiography along with conventional electrocardiography and chest roentgenography and the newer axial views of cineangiocardiography have made it possible to arrive at a precise diagnosis in defining all of the intra and extracardiac anomalies. This allows one to make full assessment of the presenting problem and offer appropriate management. Anatomical correction of severe defects is possible at present time with very little morbidity and mortality.

Key Words : *Neonate; Cyanosis; Congestive cardiac failure; Medical palliation; Surgical palliation; Surgical correction.*

Pediatric cardiology[1] has seen considerable changes in the last two decades. Changes allude to early recognition, intensive management, stabilization, and extensive use of non-invasive modalities in arriving at an amazingly high percentage of accurate diagnosis. This has evolved into offering medical and/or surgical treatment - either palliative or corrective, at an early age so as to give such an infant a better chance to have as normal a life as possible and salvage what were once thought to be "hopeless cases". Fetal diagnosis of probable, severe congenital cardiac defect(s) to alert the obstetrician, pediatrician, and pediatric cardiologist to the imminent problems at birth, so as to be appropriately prepared for such a newborn is increasing at the present. Woodrow Benson Jr. in his report to the Pediatric Cardiology Section of the American Academy of Pediatrics identified the changes in the diagnosis and treatment of congenital heart disease in the last 20 years, (a) a change from pediatric to infant and neonatal cardiology. (b) shift from palliative to more definitive surgical procedures. (c) Changes from invasive [cardiac catheterization] to non-invasive [echocardiography] diagnostic methods. (d) The surgical treatment of the most complex lesions and (e) Improved recognition. According to his report, of all children admitted to hospitals

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for treatment of congenital heart disease - those less than 3 days of age had increased from 24% in 1969-1972 to 33% in 1982-1986; and percentage of those infants with congenital heart disease who received no intervention before death had decreased from 4.7% to 1.3% in the past 10-20 years.

FETAL PHYSIOLOGY[2-4]

Complex structural abnormalities of the heart are compatible with survival to term. Normal fetal heart functions as a common blood mixing and ejection chamber which is the reason why even with complex lesions there is progress uneventfully through pregnancy. The uniform oxygenation and perfusion pressure of the fetus allows each to grow normally. It is only after birth, with the replacement of the placenta by the lung, gradual closure of the ductus arteriosus, and obliteration of foramen ovale, signs and symptoms of severe malformations begin to appear.

To understand as to why a newborn with severe cardiac malformations develops significant symptoms at such an early age and yet tolerates the same lesions and develops normally in utero, one has to be aware of the fetal circulation and the physiologic changes that occur following delivery. The three main routes in fetal life - foramen ovale, patent ductus arteriosus, and ductus venosus along with low resistance placental circulation are the ones that determine the changes. There is no question that there is high incidence of congenital heart disease in aborted fetuses, probably a reflection of extreme malformations resulting in incompatibility even in the developing fetus. Fetal and placental circulation is capable of handling any critical, so called "ductal dependent" lesions through gestation but become symptomatic shortly after birth when ductal closure occurs. Fetal and neonatal pulmonary vascularity is sensitive to changes in arterial oxygen tension augmented by hypercarbia and acidosis.

In the mature lung, the arteries accompany the airways and they develop together during fetal life. The preacinar arteries and airways [those that extend down to, but not including, the respiratory (acinar) region of the lung] are developed completely by the 16th week. Whereas, the intra-acinar arteries develop relatively late in fetal life and continue to form after birth as the alveolar ducts and alveoli develop. In the adult, the proximal half of the preacinar arterial pathway down to the 7th generation has an elastic wall structure and beyond this it is muscular. In the fetal life, this is achieved by the 19th week of gestation. In the smaller preacinar and intra-acinar arteries, wall structure is related to the diameter of an artery, the muscle giving way to a partially muscular structure and this to a non-muscular structure that is the same diameter in the fetus as in the adults lung but is located at a more proximal level along the arterial pathway. At birth, therefore, relatively few intra-acinar arteries contain muscle. Muscle cells gradually differentiate in progressively more peripheral arteries as the child grows. During fetal life, the wall thickness of the muscular and partially muscular pulmonary arteries is high and the lumen is small. As new arteries are formed, they acquire a muscle coat whose thickness is commensurate with their size. Thus, as gestation advances the number of muscular arteries rises increasing the amount of smooth muscle per unit area of lung tissue. Premature infants are born with arteries that are slightly smaller than normal and therefore have less muscle than is normal. Immediately after birth, pulmonary vascular resis-

tance falls rapidly as the lungs are ventilated and the pulmonary blood flow increases. In humans, pulmonary artery pressure falls abruptly from a mean of approximately 60 to 30 mm Hg. during first 10 hours of life and gradually achieves a normal mature level during the first month. Postnatal reduction in pulmonary vascular resistance is associated with a dramatic increase in the cross-sectional area of the pulmonary vascular bed.

INCIDENCE[5-8]

For the most part studies done so far have not shown any significant increase in the incidence of congenital heart disease at birth. Studies from several countries have a figure of 6-8 per 1000 live births, and the only indication of increase in number of new borns recognized as having congenital heart disease is because of newer modalities combined with increased awareness and perhaps involvement of pediatric cardiologists at the time of even a simple suspicion of findings to indicate cardiac problem. Perhaps the only new thing that might be accounting for higher incidence is due to increased use of drugs/alcohol in prime age mothers and possibly higher incidence of late age pregnancy with resultant chromosomal aberrations where there is a definite high incidence of congenital heart disease. Incidence will vary on the basis of the progress made in recent years. In assessing the incidence, one should take into account the following factors.[5] (i) *Population factors* such as migration, race, ethnicity with cultural, socio-economic and genetic differences including consanguinity may alter predisposition to anomalies, exposure likelihood, access to and use of medical care. (ii) *Family factors* such as maternal age, reproductive history [fetal loss, low birth weight, infant mortality] may alter the medical care sought and received. (iii) *Community medical practice* varies in availability, access and cost, suspicion of an anomaly, screening by fetal ultrasound, amniocentesis and ultimately in a physicians' decision to seek speciality advise for particular patients at various ages. (iv) *Cardiology centre practice* has also altered over time in the diagnostic definitions, in the availability and use of invasive and non-invasive diagnostic studies and in the definition of the cardiac defects. Environmental influence may also have a role in the increased birth prevalence of congenital heart disease in the communities. Study done in two areas in Arizona showed higher incidence in Yuma City [Southwestern corner of Arizona] than in Sierra Vista area [Southeast portion of Arizona]. Figures from 1983 - 1988 showed incidence of 10.5 per 1000 in Yuma compared to 5.4 per 1000 in Sierra Vista areas. Fixler reported an incidence of 6.6 per 1000 life births in Dallas County for the period 1971 - 1984. Rates for severe cases requiring cardiac catheterization or surgery or undergoing autopsy was 3.1 per 1000. Time trend for rates of congenital heart disease suggested an apparent increase in prevalence rate during the 1970's which could be accounted for by an increase in detection of mild cases as reflected by a greater tendency for pediatrician to refer asymptomatic children with significant heart murmurs to a pediatric cardiologist. Similar prospective analysis done in Czechoslovakia by Simonic *et al* showed a prevalence of 8.223 per 1000 live births.

ETIOLOGY[9-11]

Malformations may be caused by gene defects, chromosome abnormalities, and environmental factors, the interaction of

many genes in the environment in multifactorial etiology, or maternal cytoplasmic transmissions of mitochondrial DNA in mitochondrial diseases. Congenital heart defects manifest etiologic heterogeneity although most cases are as a result of multifactorial etiology. Stronger genetic influences are now being recognized.

Ninety percent of all congenital cardiac malformations have a genetic - environmental interaction [multifactorial inheritance], 8% are accountable on the basis of primary genetic factors [chromosome 5%, single mutant gene 3%]. Primary environmental factors like for example Rubella and others are seen in the remaining 2%.

Probable cardiac teratogens include alcohol, amphetamines, hydantoin, lithium, retinoic acid, thallidomide, rubella, maternal diabetes and maternal phenylketonuria.

Anatomical completion of the cardiac structures occurs within the first trimester of pregnancy. Presumed vulnerability period for teratogenic influence on the cardiovascular development is between 18 to 60 days gestation as outlined in Table 1.

RECOGNITION[12-15]

There are relatively few ways in which an infant with cardiac disorders presents during the neonatal period. High index of suspicion needs to be there to identify these infants. Obvious manifestations vary depending on the type of congenital heart defect the infant has. Apgar score still has it's value in predicting the course of the infant at birth. Low score due to persisting cyanosis, dropping heart rate or respiratory distress may or may not be related to underlying cardiac pathology but one that should be kept in mind and ruled out by appropriate tests. Although certain cardiac lesions do not manifest themselves [cyanotic/ obstructive/shunt lesions] perhaps for a few days to a few weeks after birth, while examining a neonate for congenital cardiac defects one should look at other systems as certain syndromes could be present that might give us a clue. Conditions like ear lobe anomalies, abnormal fingers, simian crease, absent of finger-like thumb, radial hypoplasia, hypertelorism, slanting eyes, long philtrum with uptilted nose, etc., would all indicate certain syndromes to be present which are known to be associated with intracardiac anomalies. Any infant noted to have multiple system involvement should be followed closely for any evidence of con-

TABLE 1. Teratogenic Influence and Period of Vulnerability

Abnormality	Embryonic event completed	Limits of vulnerable period	Most sensitive (Day of Development) vulnerable period
Conotruncal	34	14-34	18-29
Endocardial cushions	38	1438	1833
Ventricular septum	38-44	14-44	18-39
Atrial septum (secundum)	55	14-55	18-50
Semilunar valves	55	14-?	18-50

genital heart disease. Any concern in palpating liver as to whether it is left sided or appears to be midline should lead one to suspect the possibility of polysplenia or asplenia syndrome.

Manifestations of underlying cardiac pathology are primarily seen in the neonate as : cyanosis, congestive heart failure, abnormal heart rate and cardiac murmur.

A. Cyanosis

Cyanosis in the newborn and infant although not synonymous with "arterial desaturation" is recognized as indicative of serious problem. Often perceived as an ominous sign, in most cases it is associated with severe intracardiac malformations. Neonate with cyanosis causes considerable concern with the physicians caring for the infant and poses a challenge to delineate the exact cause so as to institute appropriate treatment to prevent any complications. Although following the first few minutes of life, central cyanosis often indicates congenital heart disease, alternatively, right to left shunting through a patent foramen ovale or patent ductus arteriosus in the absence of other cardiac defects can lead to marked central cyanosis in those infants with persistent pulmonary hypertension with or without associated pulmonary parencymal disease. Even in the absence of pulmonary hypertension, central cyanosis secondary to right to left shunting through it's persistent fetal pathways can be noted intermittently particularly with crying through the first several days of life. The major problem one considers and makes every effort to identify with all the diagnostic modalities available is cyanotic congenital heart disease. The seriousness and urgency in recognizing such a problem lies in the fact not to take the infant for diagnostic studies but to stabilize the infant first.

Cyanosis is either central or peripheral. Central cyanosis relates to the presence of reduced hemoglobin of whatever etiology. It is recognizable when more than 3 g/dl of reduced hemoglobin is present. Clinical central cyanosis becomes apparent when oxygen saturation varies from about 62% when the hemoglobin is 8 g percent to about 88% when hemoglobin is 24 g. percent. Thus, anemia mask visible cyanosis and polycythemia accentuates it. Relative polycythemia in the newborn [14-20 g/dl] facilitates early and easy recognition. However, presence of fetal hemoglobin makes it more difficult since fetal hemoglobin has more affinity for oxygen. For example, at a given partial pressure of oxygen, fetal hemoglobin is more saturated than adult hemoglobin. Causes of central cyanosis are listed in Table 2.

Peripheral cyanosis characterized by cyanotic extremities (normal PAO2 and oxygen saturation) is common in the neonate and persists for hours to weeks (acrocyanosis). Such discoloration usually disappears by 48 hours and is rarely seen after 72 hours. Vasoconstriction associated with cold or acidosis prolongs or precipitates acrocyanosis. It is usually related to vasomotor instability that causes peripheral vasodilation with resultant venous stasis and increased oxygen extraction by the tissues. Mottling of the skin is an example of such a phenomenon.

Approximately one-third of neonates initially suspected of congenital heart disease are found to have no structural congenital cardiac abnormalities when the evaluation is completed. Of the 6-8 per 1000 live births 30% belong to cyanotic heart disease category.

TABLE 2. Causes of Central Cyanosis

1.	Congenital heart disease - right to left shunt and decreased pulmonary blood flow;
2.	Congenital heart disease - right to left shunt with increased pulmonary blood flow;
3.	Congenital heart disease with alveolar hypo-ventilation due to heart failure;
4.	Primary lung disease;
5.	Mechanical interference with lung function;
6.	Persistent fetal circulation syndrome;
7.	Central nervous system disease;
8.	Hemoglobinopathy;
9.	Hypoglycemia;
10.	Polycythemia;
11.	Shock and sepsis; and
12.	Miscellaneous [neuromuscular disorders, maternal medication, etc.]

Skin color as an indicator of state of oxygenation can, however, be complicated by a number of conditions unrelated to oxygen saturation. Nursery lighting, wall paint, skin pigmentation and even clothing are well known examples of giving false impression of cyanosis.

True central cyanosis with hypoxemia can most easily be distinguished by examination of the tongue or oral mucosa which are not involved in peripheral cyanosis. Although improvement of color with oxygen administration may lead one to think of non-cyanotic cardiac lesions, the best way at least at present, is to measure partial pressure of oxygen (PO_2) in room air and in 100% oxygen for evidence of true cyanosis. Since most institutions now have capabilities of using umbilical artery catheters direct arterial sampling is possible, if not, peripheral arterial sampling could be done easily. Transcutaneous partial pressure of oxygen measurement monitors - under certain conditions and reservations are also used. Pulse Oximeter to a certain extent has its value in measuring oxygen saturation directly. In spite of all these gadgets, one should keep in mind that unless one is dealing with significantly hypoperfused lungs or parallel circulation as in transposition, PO2 measurements by transcutaneous monitor and oxygen saturation by pulseoximeter may not rule out cyanotic congenital heart disease.

Cyanosis is a serious sign in a newborn and one to be treated with appropriate respect. Cyanosis persisting after a few hours in oxygen in an infant not previously in respiratory distress should never continue to be "just observed". Major mistakes occur because cyanosis is dismissed lightly at this age. Early diagnostic study in such an infant under stable conditions before any deterioration and/or acidosis developing thereby complicating the picture, is well tolerated and gives useful data for an accurate diagnosis and management either medically and/or surgically.

Unusual distribution of color changes may be explained by massive pulmonary - aortic shunts occurring through the ductus arteriosus. The upper part of the body will then be a different color from the lower half, the line of demarcation usually being just below the umbilicus. Occasionally the left arm becomes the same color as the lower compartment under these circumstances. Most often this change is represented clinically by a pink upper and cyanotic lower division indicating intact septa. Usually this is the result of a transient pulmonary disorder producing critically high pulmonary vascular resistance and temporary right to left ductal shunt. Less often obstructive lesions in the left heart can be held responsible for such color difference. For

example, severe coarctation of the aorta or interruption of the aortic arch with ductal right to left shunt will give cyanosis of lower half of the body. The frequent association of a ventricular septal defect permits arterialized blood to enter the pulmonary artery, therefore the descending aorta so that the arterial oxygen saturation differences between ascending and descending aorta will be small. The contrasting situation of pink lower extremity and cyanotic upper body is seen when coarctation of aorta and patent ductus is present with complete transposition of the great vessels.

Conditions causing marked cyanosis are : (5T and E) Transposition of great arteries, tertralogy of Fallot, tricky pulmonary valve (pulmonary valve atresia or critical, pulmonic stenosis with intact ventricular septum), tricuspid atresia, total anomalous pulmonary venous connection (obstructive type), Ebstein malformation of tricuspid valve. (Reference 12. p. 143).

B. Congestive Heart Failure

With the improved knowledge and early recognition of congenital cardiac malformations, advanced congestive heart failure is less frequent nowadays. Yet, under certain conditions, there is a possibility of seeing an infant who comes with severe congestive cardiac failure with extreme collapse as is often seen with coarctation of aorta syndrome wherein the ductus closes abruptly and causes severe left heart failure. The early warnings of congestive failure are increased respiratory rate, tachycardia or gallop, loud heart sounds, large liver, abnormal pulses and questionable cyanosis (Table 3).

In the newborn, congestive heart failure presents as a clinical syndrome involving all systems. The clinical manifestations of the pumps' inability to dispose adequately of venous return and/or supply the body with cardiac output demands can be quite variable. Additionally, myocardial reserve in the newborn is minimal and very rapid progression of congestive heart failure is not uncommon. Cardiac failure presenting at birth is rare and usually a manifestation of hydrops fetalis associated with intravascular hemolysis, invariably present in infants with congestive heart failure but neither the heart rate nor the respiratory rate exceeds values that are accepted as upper limits of normal. Rather the typcial finding is persistence of values near the upper end of the normal range, with loss of normal variability. Gallop rhythm may or may not be present, but when present it generally indicates more advanced dysfunction. Pallor with peripheral vaso-constriction is invariably present and is often the most striking early finding particularly in premature infants. Pulses are poorly felt and the pulse pressure is

TABLE 3. Guidelines for Defining the Presence of Congestive Heart Failure in the Newborn Infant

A.	*Borderline* congestive heart failure.
	Any three of the following:
	Cardiomegaly (cardiothoracic ratio 60)
	Tachycardia (150/min.)
	Tachypnea (60/min.)
	Wet lungs
B.	*Moderate* congestive heart failure.
	Satisfaction of Criteria for A. plus any one of the following:
	Gallop rhythm
	Hepatomegaly (3 cm. or more)
	Frank pulmonary edema
C.	*Severe* congestive heart failure.
	Vascular collapse.

(Reference 12. p. 157).

narrow even in infants in whom failure is the result of systemic run off into a lower resistance circulation. Unusual weight gain is also common although somewhat of a later finding, and peripheral edema is rarely seen. Hepatomegaly is a late finding and splenomegaly is almost never noted. Cyanosis and rales indicate overt pulmonary edema and are seen in advanced myocardial failure.

It is important to recall the findings of congestive heart failure will mask clinical ausculatory findings that might permit accurate elucidation of the underlying lesion responsible for the development of myocardial dysfunction as murmur(s) may be absent or poorly heard. Accordingly, appropriate treatment of congestive heart failure may bring out true features that would be typical for the diagnosis. Interpretation of the findings observed in the presence of congestive heart failure should be performed with great caution. Frequently diagnosis of underlying lesion that precipitated myocardial failure must either wait resolution of congestive heart failure or be carried out using other methods other than simple auscultation.

Conditions causing congestive heart failure :

Patent ductus arteriosus; hypoplastic left heart syndromes; coarctation of aorta syndrome (including aortic arch interruption); ventricular septal defect; endocardial cushion defect; truncus arteriosus; total anomolus pulmonary venus drainage (non-obstructed type); aortic stenosis; double outlet right ventricle; and arteriovenous fistulae (Reference 12, p 143)

C. Respiratory Activity

In newborn infants the work of breathing is done principally with the diaphragms so that respiratory movement or excursion is most easily judged from the lateral view of the upper abdomen and chest. In the first hours of life the average respiratory rate is about 60 breathes per minute. Transient variation from 20 to 100 breathes a minute can be seen occasionally. Between 2 and 6 hours after birth, average rate decreases to 50 breathes per minute (range 20 to 80) and then to 30 to 40 breathes per minute (range 20 to 60) after six hours. Throughout the first 12 to 24 hours of life breathing is more often irregular than regular, and recurrent, brief, less than 3 second pauses are common. Most infants with heart disease will not have respiratory rates outside these ranges. They will, however, often maintain a rate that is consistently near the upper end of the normal range. Additionally, the normal irregularity of the breathing pattern may be lost. Infants with diminished pulmonary blood flow or with transposition of the great vessels will most often demonstrate a rapid shallow respiratory pattern. The rate persistently varies between 50 and 60 beats per minute for term infants, between 60 to 80 beats per minute in pre-term infants. The rate will often increase rather than decrease with increasing age. Respirations will however still be performed utilizing the diaphragm so that rapid shallow movements of the abdominal wall may be the most striking feature on observation. In contrast, any infant with pulmonary edema, pulmonary parenchymal disease or airway obstruction, tachypnea will be accompanied by a pattern of irregular respiratory activity with deeper breathing and intercostal and subcostal retractions.

D. Heart Rate

Resting rate averages 120 to 130 beats per

minute the first week of life in the term neonate increasing to 150 beats per minute during the second to fourth weeks. During the first week the resting rates below 100 beats per minute or above 160 beats per minute is present in less than 5% of the normal term infants. Transient fluctuations in rate as high as 220 beats per minute or as low as 59 beats per minute have however been reported to occur normally in healthy term newborns during the first three days of life. Bradycardia is generally not an indication of underlying cardiac disease.

Arterial pulse and blood pressure. Simultaneous branchial and femoral pulse needs to be palpated. Normally they are synchronized and have an equal excursion to the fingers. Best assessment is made with the infant warm and if not asleep, at least comfortable and relaxed. With brisk branchial and weak or delayed, or impalpable femoral pulse cause is most often coarctation but may be aortic stenosis, supravalve aortic stenosis or congenital hip dislocation. Occasionally coarctation may not manifest for a few days because of aortic end of ductus staying open and bypassing the obstruction. Brisk jerking or collapsing pulse is seen in patent ductus arteriosus, AV malformation, congestive heart failure secondary to ventricular septal defect. Absent pulse is seen in generalized vascular collapse of advanced congestive heart failure in neonates. Rarely in coarctation, left branchial artery may be brisk with decreased or absent right branchial that arises from the descending aorta below the coarctation.

Many centers have Doppler techniques that have replaced the traditional blood pressure measurement tools. Doppler principal allows direct detection of blood flow by the ultrasonic frequency shift produced by moving blood particles. A cuff and sphygmomanometer applied in the usual manner, the blood flow is detected by an ultrasonic probe placed over the artery distal to the cuff. As with standard methods of measurement, cuff size is very important. The cuff bladder should encircle the extremity and be the widest that can be applied at the upper arm (or leg). For the term neonate this generally means that a 4 to 5 cm. cuff should be used. Doppler method gives only systolic pressure as opposed to indwelling catheters where both systolic and diastolic pressures are measured. Normal systemic blood pressure in full term neonates averages 70 to 70 mmHg. (range 55 to 90). Doppler systolic blood pressure obtained are similar averaging 66 mm Hg. Blood pressure in infants can be greatly affected by body temperature, activity and posture. Pressures obtained in the leg are generally slightly higher than those in the arm but can be equal or slightly lower. The systolic pressure in the upper extremities if more than 20 mmHg higher than that in the lower extremities is highly suggestive of aortic coarctation. It should be noted, however, that in the presence of a patent ductus arteriosus that permits free passage of blood around the coarctation area, the pressure difference in the coarctation may be masked. In cases of interrupted aortic arch, aortic coarctation involving the left subclavian artery or coarctation with aberrant right subclavian artery there may be differential pressure between the arms as well as between the upper and lower extremities.

E. Cardiac Murmur

Sixty percent of healthy babies have innocent heart murmurs. Patent ductus arteriosus either classical continuous type or

abbreviated type continuous murmur or crescendo systolic murmur may be heard in 14% of healthy term infants somewhere between birth and up to 15 hours of age. Any interference with normal transitional circulation can influence the ausculatory findings markedly. Ejection systolic murmurs of vibratory quality in the second left intercostal space and Grade I-II/VI in intensity can be heard in almost 60% of infants in the first few hours of life. These may be occasionally heard in the back also related to pulmonary artery bifurcation. Presence of a Grade III/VI or more heart murmur, palpable thrill, single second heart sound beyond 12 hours of age, ejection click beyond 12 hours of age are considered as abnormal ausculatory findings.

ELECTROCARDIOGRAPHY

In assessing the electrocardiogram one has to keep in mind the "normal" right ventricular dominance that is seen up to a week of age (upright T-waves and prominent R-wave in V-1). Any neonate whose electrocardiogram shows prominent R-wave exceeding 21 mm. in V-1, pure R-wave in V4R and V-1, or QR pattern in V4R and V-1 or abnormal RS progression to indicate increased left ventricular forces should be evaluated for possible congenital heart defects. Presence of right atrial enlargement leads one to suspect right sided lesions and occasionally left sided obstructive lesions (critical aortic stenosis with LV dysfunction as in the case of coarctation syndrome). Normal axis of 180° is noted in newborns (+ 30 to + 180°). Any variation, especially with axis in the northwest corner (+180 to +270°) or left axis deviation is to be considered abnormal and all attempts should be made to rule out or identify the cardiac lesion. Certain electrocardiographic changes are very diagnostic. For example, in a cyanotic infant, left axis deviation with left ventricular hypertrophy should lead one to suspect tricuspid atresia unless proven otherwise. Rightward axis with left ventricular hypertrophy and right atrial enlargement might indicate pulmonary valve atresia/critical pulmonic stenosis with hypoplastic right ventricle. Right axis deviation with severe right ventricular hypertrophy with very poor left ventricular voltage may mean hypoplastic left heart syndrome. Likewise, presence of ventricular pre-excitation of type-B in a cyanotic infant with enlarged heart might lead one to suspect Ebstein's malformation of tricuspid valve.

CHEST ROENTGENOGRAPHY

As part of workup in a neonate suspected of congenital heart disease, chest roentgenography, certainly has its own merits. In most instances a PA and lateral view is obtained. Proper positioning and well exposed film as well as exposing the film during full inspiration adds significantly to diagnostic capabilities. Cardiac size, shape, position of the heart, side of aortic arch, lung vascularity, abdominal viscera and lastly any demonstrable skeletal anomalies that are visible should be assessed. Keep in mind that a normal heat size does not rule out congenital heart disease nor an enlarged heart rules in congenital heart disease as seen in infants with nonstructural heart disease secondary to a diabetic mother, perinatal hypoxia and its effect on myocardium. Certain typical findings are nearly diagnostic. An "egg" shaped heart with its rounded apex, narrow base is seen in transposition of the great arteries; gross enlargement of the heart with very prominent right atrium as in pulmonary atresia with intact ventricular septum, enlarged heart with rounded apex and

increased vascular markings as in hypoplastic left heart syndrome; normal heat with "lacy type" vascular markings or "cotton wool" appearance of venous congestion as in total anomalous pulmonary venous connection of the obstructed variety, are some of the examples. Hypovascular lung fields mean decreased pulmonary blood flow with right-to-left shunt as seen in tetralogy of Fallot, pulmonary atresia, tricuspid atresia with associated pulmonary stenosis and normal great vessel relationship as well as Ebstein's anomaly. Increased vascularity with cyanosis may be related to total anomalous pulmonary venous connection without obstruction, hypoplastic left heart syndrome, coarctation syndrome with some left ventricular dysfunction and atrial left-to-right shunt. Isolated dextrocardia with situs solitus in a cyanotic infant or findings to suggest abdominal heterotaxy with dextrocardia leads one to suspect asplenia/polysplenia syndrome both of which are associated with severe multiple congenital cardiac defects. Right aortic arch is commonly associated with tetralogy of Fallot and truncus arteriosus.

ECHOCARDIOGRAPHY[16-20]

Precise anatomic diagnosis has become a reality with the advent of echocardiography. With improved technology, and added color flow and Doppler techniques, identification of cardiac anatomy and associated pathology has become almost as diagnostic as cardiac catheterization. Under certain conditions, with sick neonates, some centres routinely send patients to surgery without the benefit of cardiac catheterization. At times it may be necessary to mildly sedate the infant to obtain all necessary information. A good technician with the help of a pediatric cardiologist guiding the technician, a thorough study is possible. Along with the conventional long axis, short axis, four chamber (apical and subcostal) as well as aortic arch studies, sometimes unorthodox views may be required to identify the anatomy and direct supervision and guidance of the pediatric cardiologist caring for the neonate, who wants to get as much information as possible, is essential. The well standardized segmental analysis as an approach to identify anatomical structures, to know the relationship, and to interpret color flow studies along with Doppler interrogation of all the valves as well as descending aorta and including any areas where abnormal color flow due to increased turbulence is present, it is essential for a pediatric cardiologist to be physically present at the time of the study in such a sick neonate with suspected congenital heart disease. This has yielded excellent results to the point of sending these neonates to surgery without the benefit of cardiac catheterization.

Limitations in getting an adequate study are due to three major factors : (1) *Patient* - crying, nursing infant or sitting up often leads to errors or omissions. Infants should be quite and supine. If necessary using mild sedation (Chloral hydrate) would help. (2) *Echocardiographer* who should have the basic knowledge of cardiac anatomy and pathology of congenital cardiac defects and must be experienced in the examination of infants. (3) *Equipment* - should have excellent near field resolution and high frequency transducers for good lateral resolution, the machine itself should be easily portable and small enough to use in the intensive care isolette.

CARDIAC CATHETERIZATION[13, 21-25]

Inspite of the ever increasing belief that

echocardiography is capable of giving all necessary information to make a therapeutic decision, there are times when one ought to consider further diagnostic invasive study. This, especially becomes important under certain circumstances if one is contemplating on interventional procedures. Even here, some institutions are, at least capable of doing these under echocardiography. Although one likes to keep away from stressing a neonate and avoid associated morbidity and perhaps even the slightest mortality that has been reported, invasive study has to be considered to assess the exact anatomical defect(s), physiologic consequences and to make a decision as to whether palliative or total repair could be attempted.

With due consideration given to acid-base balance, metabolic state, stabilizing blood sugar, attending to temperature control and using pulse oximeter, even under conditions of intubation and ventilation of an infant, this has to be done. Although conventional femoral venous approach has been the usual method of arterial as well as venous entry, umbilical vessels have also been used for not only venous but also arterial catheterization studies. Using a non-ionic contrast material and obtaining biplane Cineangiograms would minimize the effect of more dye as well as getting more information with the biplane technique than in the single plane technique. Again, using various axial views would certainly give better anatomical details. One should also keep in mind that only the information required to arrive at the diagnosis is to be done in a critically ill infant rather than prolonging the study and making the neonate run into difficulty. Warming the room (temperature control), using a heating pad under the infant, presence of oxygen source nearby along with suction equipment and facility to intubate and ventilate are essentials in a pediatric cardiac catheterization laboratory as also trained pediatric personnel.

As mentioned earlier, under certain conditions, during cardiac catheterization palliative procedures could be carried out. Balloon atrial septostomy as in transposition of the great arteries has been a lifesaving measure even now in some infants with restricted atrial communication as seen in certain right or left sided obstructive lesions that would require decompressive mechanism. Palliative valvuloplasty - for example, pulmonary valvuloplasty in severe tetralogy of Fallot or critical pulmoic stenosis has been successfully carried out so as to allow the infant to become better saturated and avoid emergency surgery. Of course, in those with stenotic lesions like pulmonic stenosis, aortic stenosis, coarctation of the aorta and pulmonary arteries, balloon valvuloplasty and angioplasty have become treatments of choice thus avoiding surgery although one needs to keep complications in mind.

MANAGEMENT[26-32]

General. As alluded to earlier in this paper, initial management of a sick neonate with congenital cardiac lesion(s) is of utmost importance if one has to avoid any complications developing later. In those infants who are having respiratory compromise, endotracheal intubation and mechanical ventilation should be done promptly. Acid-base status assessed as often as necessary to avoid metabolic acidosis or alkolosis which in turn would avoid any myocardial dysfunction should be treated promptly. Adequate oxygenation to keep a satisfactory satura-

tion again to prevent any hypoxaemia and acidosis is to be followed. Hypocalcemia, hypoglycemia that are often common in these sick neonates, especially cyanotic neonates, should be attended to. Temperature regulation to prevent hypothermia which in turn could give rise to severe vascular compromise and perhaps abnormal metabolic process and acidosis during evaluation and diagnostic studies should be carefully attended to. Fluid balance along with adequate caloric intake with high glucose concentration, total parenteral nutrition if necessary, and/or feeding, even if one has to resort to tube feedings, is part of the overall management. If able to nurse or bottle feed, and if condition does permit, it should be encouraged.

Medical. With the elucidation of prostaglandins, it has been a liefsaving measure in those infants who have ductal dependent cardiac lesions to start on prostaglandin-E infusion to keep the ductus open thereby allowing the infant to become more stable and avoid emergency measures including surgery at least in some of them. Things could be "planned" for such an infant once the neonate becomes stable on prostaglandin and pediatric cardiologists think this as a "luxury" on hand. Complications relating to the use of prostaglandin include temperature elevation, increasing white cell count with left shift that may mimic sepsis, as well as platelet dysfunction that might cause some bleeding problems. Apneas are well documented with prostaglandin infusion and one should be aware of this and may need to intubate the infant if necessary. Prostaglandin-E in the dose of 0.05-0.1 microgram/kg is used to keep the ductus open and titrated to keep the oxygen saturation at a decent level.

Hypotensive episodes due to "poor pump" are treated with the use of Dopamine or Doubutamine infusions and if there is hypertensive crisis as in coarctation of the aorta, use of anti-hypertensive drugs, (vasodilators and/or ACE inhibitors) is essential to improve failure.

Conventional methods of medical management of congestive heart failure still hold good in the present era in spite of all the developments one has seen. Digitalis along with use of diuretics and controlling fluid intake is the standard rule still applied to control congestive cardiac failure in these neonates.

Surgery. Although a sick neonate is looked upon as one who carries higher morbidity and mortality for surgical therapy of underlying cardiac defect, recent improvements have made this procedure possible with significantly reduced morbidity and mortality. The progress made in surgical treatment of congenital heart defects in the last two decades has been tremendous. With improved instrumentation, techniques have been applied to neonates not only to palliate but also to correct those that are amenable to such a treatment once thought to have very high morbidity and mortality. Development of microvascular techniques, microvascular instrumentation, routine use of systemic hypothermia to levels less than 20° centigrade with or without circulatory arrest, ventilatory management of the neonate and the effects of prior stabilization of a sick neonate in attending to oxygen requirement, metabolic abnormalities, temperature regulation as well as advent of therapeutic manoeuvers like use of prostaglandin has improved the outcome tremendously. Modern techniques of anaesthesia without depressing cardiac function has also been recognized as a major step towards these goals. Primary repair of intercardiac defects

not only of the left to right shunts but also of the cyanotic lesions (tetralogy, transposition, etc.) along with correction of anomalies of the venous return as well as critical stenotic lesions has been done with very little morbidity and mortality.

Surgical palliation. Initially, palliative procedures were performed because corrective surgery was not available or carried a high risk. As operative correction of more complex cardiac lesions became feasible, palliation was used primarily in infants since all children deemed too small or too ill to undergo correction. Currently, with corrective open heart surgery of even very complex lesions being performed in neonates in many centres, the role of palliative surgery for congenital heart disease is being further restricted. Palliation is used in lesions in which correction is not feasible in the neonatal period but is an operation that requires low pulmonary vascular resistance such as aortopulmonary anastomosis, operations that require use of prosthetic tubular grafts since these will require replacement as the child grows and in children who are too ill or weak from malnutrition or sepsis to undergo an open heart operation. Palliative surgery is also used when the risk of corrective procedure decreases rapidly with age. Lesions that fall into this category include tetralogy of Fallot, atrio-ventricular septal defects with right ventricular outflow obstruction or multiple ventricular septal defects. As the operative techniques of repair and post operative care of these infants improve, the indications for palliation continue to decrease. Palliative operations can be subdivided into three major categories (*a*) *Increase pulmonary blood flow* is indicated in those where there is decreased pulmonary blood flow. Shunts used are the classic Blalock-Taussig, modified blalock-Taussig and enlarging right ventricular outflow tract. Because of their inherent complications, Waterston and Pott's shunts are not used presently. (*b*) *To decrease pulmonary blood flow* - Pulmonary artery banding. (*c*) *To improve venous mixing* - Atrial septectomy. (*d*) *Balloon valvuloplasty angioplasty* of severe stenotic lesions.

Pulmonary artery banding is a useful palliative procedure for a diverse group of patients with congenital cardiac anomalies and unrestricted pulmonary blood flow. With improved results of primary repair of intracardiac anomalies in small infants however, pulmonary artery banding should be reserved for severely ill infants with complex lesions not amenable to early difficult correction. Currently, pulmonary artery banding is indicated in patients with excessive pulmonary blood flow and single ventricle or tricuspid atresia. Pulmonary artery banding is also appropriate in certain patients with atrio-ventricular communis defects and in patients with muscular multiple ventricular septal defects. Pulmonary artery banding is an option in patients with ventricular septal defect and coarctation of the aorta although more and more centres are now going for one stage correction through mid-line sternotomy incision.

Total correction. As mentioned earlier, with the advances made and the improved anaesthesia, hypothermic cardiac arrest and micro-surgical tools along with the excellent surgical techniques, more and more centres are doing total corrections of congenital cardiac defects - not only simple ones but also complicated ones with remarkably low morbidity and mortality. Improved intensive care management in the immediate period and post operative period as well as

further understanding of pathophysiology, pharmacology of drugs, recognition of various rhythm disturbances with therapy appropriately directed to control such rhythm disturbances, the stride one has taken is unimaginable.

It is beyond this paper to go to the details of the surgical approach and excellent references are available for one to review.

Lastly, I would like to present here a brief note on the premature infant with patent ductus arteriosus complicating hyaline membrane disease.[33] One should be aware of the complication of patent ductus arteriosus in a premature infant who develops significant pulmonary problems with increasing oxygen requirement and continued ventilatory dependency, eventually developing serious complications of bronchopulmonary dysplasia. Under these circumstances, once patent ductus arteriosus is recongized, intervention is advised to avoid any serious late complications. It has been well established that in those premature infants under 1250 gms. therapeutic intervention of patent ductus arteriosus, although does not have long term beneficial effects, has shown by several studies, at least to prevent increasing oxygen requirement and prolonged ventilator dependency, hence either medical or surgical palliation is done. Medical palliation in these infants is the use of Indomethacin to prevent prostaglandin synthesis in the ductal smooth muscle, thereby allowing the ductal smooth muscle to constrict and decrease the shunt or even close completely. Intravenous Indomethacin has been used widely in a dose of 0.2 mg/kg as an initial dose and subsequent two dose twelve hours apart at 0.1 mg/kg. Careful management of fluid intake and attending to renal function to make sure that there is no acute renal complications, as well as any hematological complications in the form of bleeding tendency and possible vascular spasm in the intestinal mucosa which eventually might lead to necrotising enteroscolitis are to be kept in mind. Initial laboratory investigation should include clotting factor studies along with bilirubin as well as blood ureas nitrogen and creatinine determination and also possibly use of ultrasound of the head for any evidence of bleeding before Indomethacin is given and subsequent repeat study to make sure that no such complication is seen. Studies from Vanderbilt University have shown improved success in these infants. If there is any contraindication for Indomethacin use, surgical ligation of the ductus is recommended and recently almost all centres with intensive care nurseries, this procedure is being carried out without any undo-complications in the nursery itself thus avoiding risks associated in transporting such sick neonates. Symptomatic patent ductus arteriosus is reported in 45% of pre-terms under 1 kg. birth weight, up to 28% between 1 and 1.5 kg., and less than 10% in infants over 1.5 kg. In a randomized clinical trial conducted in Vanderbilt Newborn Intensive Care Unit, of the 32 patients with a birth weight between 750 and 1500 g. with respiratory distress syndrome requiring mechanical ventilation at 24 hours postnatal age, a group of 15 received a single intravenous dose of Indomethacin, .2 mg/kg at 24 hours postnatal age and control group of 17 did not receive Indomethacin. At the conclusion of the trial, it was apparent that early Indomethacin significantly reduced the incidence of symptomatic PDA. Excluding one patient in each group whose early death may have pre-empted occurrence of symptomatic PDA, 1 of 14 early Indomethacin patients and 9 of 16 controlled patients developed

symptomatic PDA. The authors believe that although neither short nor long term reductions in pulmonary morbidity have been demonstrated in clinical trials, prevention of symptomatic PDA per say is of sufficient benefit to warrant the early administration of Indomethacin to patients with high probability of developing symptomatic PDA particularly if they have RDS. For this reason, infants in the Vanderbilt NICU who weigh less than 1250 g. at birth and who require mechanical ventilation at 24 hours postnatal age are given a single injection of Indomethacin to prevent symptomatic PDA.

In summary, this article presents an overview of the neonate with congenital heart disease. The stride in the last 20 years is like what U.S. astronaut Neil Armstrong said when he landed on the moon - "A small step for man and a giant leap for mankind". An enormous array of new developments has made it possible to salvage neonates with critical as well as complicated congenital cardiac lesions. It is a matter of being aware of various manifestations these neonates present with and careful observation and a strong suspicion that leads one to identify, diagnose, and offer help to make them survive and lead a life that would not have been possible in the past but for the progress made.

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