Folia Medica Indonesiana Vol. 45 No. 2 April – June 2009 : 145-154

Review Article : CONGENITAL HEART DISEASE The Holistic Approach, Now and in The Future in Indonesia

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ABSTRACT

Medicine is an ever-changing science. As new research and clinical experience broaden our knowledge, changes in basic to clinical approach, treatment and drug therapy are required. Health and Medicine emphasizes holistic approaches to understanding and preventing disease. A wide range of health care interventions is explored. Congenital heart disease (CHD) is cardiovascular malformations are generally the result of aberrant development of a normal structure in the fetus, or failure of such a structure to progress beyond an early stage of embryonic or fetal development. Malformations are due to complex multifactor genetic and environmental causes. The only possible definitive treatment is cardiac corrective surgery. Now, in Indonesia, this procedure remains less accessible, so that a large proportion (95%) of CHD patients have no opportunity to undergo operation, subjecting those patients to the risk of fatal complications and chronic stress. Although human genetic approaches are important in understanding of the etiology of congenital heart disease (CHD), detailed molecular analysis of cardiac development in human is difficult. Recently, researchers have begun to clarify how stress may contribute to these pregnancy outcomes. High levels of stress were more likely to have high levels of a hormone called corticotrophin-releasing hormone (CRH) in their blood. This and other studies have found a link between high levels of CRH and preterm labor or congenital hear defect of the baby. A thoughtful and rational holistic approach to the prevention of the occurrence of CHD, and differential diagnosis is important for prompt recognition and appropriate management. It is imperative that a concerted team approach involving psychiatrist, geneticist, neonatologist, cardiologist, nurse, surgeon, and anesthesiologist is utilized. This paper suggests that psychoneuroimmunology may have broad implications for the basic biological sciences and medicine in CHD.

Key Words : *congenital heart disease, psychoneuroimmunology , holistic approach*

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INTRODUCTION

Heart defects are among the most common birth defects, and are the leading cause of birth defect-related deaths. However, advances in diagnosis and surgical treatment over the past 40 years have led to dramatic increases in survival for children with serious heart defects in Indonesia. The cause of most CHD is unknown. At present, the complex genetics and inheritance of CHD remains incompletely understood. In the past, the situation was even more unclear because many children with CHD did not survive to reproductive age and fetal echocardiography was not available. In most cases, it is of a multifactor origin and is a result of both genetic predisposition and environmental factors. Known genetic causes of heart disease includes chromosomal abnormalities such as trisomies 21, 13, and 18, as well as a range of newly recognized genetic point mutations, point deletions and other genetic abnormalities as seen in syndromes such as CATCH 22, familial ASD with heart block, Alagille syndrome, Noonan syndrome, and many more. Known ante-natal environmental factors include maternal infections (Rubella), drugs (alcohol, hydantoin, lithium and thalidomide) and maternal illness (diabetes mellitus, phenylketonuria, and systemic lupus erythematosus) (Anderson et al 1987; Baldwinn 2001; Sullivan 2002).

A number of studies have suggested that high levels of stress in pregnant women may increase the risk of preterm labor and low birth weight. Other findings have indicated that more premature and small-for-date babies or CHD are born to mothers who experienced severe stress, particularly during their first trimester. This pattern of responses indicates that an advanced pregnancy may somewhat protect women from the

psychological effects of acute stress and maternal anxiety is relative indications for fetal echocardiography. Again, these studies often are fraught with recall bias. In many studies where researchers prevented recall bias by asking the patients about stress before pregnancy complications occurred, findings *did not* show a greater likelihood of complications in women who experienced critical life events. Recently, researchers have begun to clarify how stress may contribute to these pregnancy outcomes.

High levels of stress were more likely to have high levels of a hormone called corticotrophin-releasing hormone (CRH) in their blood. This and other studies have found a link between high levels of CRH and preterm labor or congenital hear defect of the baby. The researchers speculate that increased levels of stress-related hormones may affect both maternal blood pressure and fetal growth and development. While this study is preliminary, it may lead to a new approach to identifying a group of women who are at risk of preterm labor and of having congenital heart defect or a low-birth weight baby. It appears that CRH or other stress-related hormones may constrict blood flow to the placenta, so the fetus may not receive the nutrients and oxygen it needs for optimal growth (Artman et al 2002a, Anderson et al 1987; Dinarrevic et al 2000).

Stress results in the perception of emotional strain as well as physical responses, such as an increase in heart rate or stress hormones. Research on stress can be done in a number of ways: by asking subjects about their perceived level of stress, by actually measuring physical markers of stress, or by taking both factors into account. Study results on maternal stress during pregnancy is also found to cause cardiac embryonic disturbances of the fetal, even though the effects of stress on pregnancy outcome have been conflicting. Some findings have shown that during the first trimester and postpartum stress is felt more deeply, or at least has a greater effect on physical factors (Artman et al 2002a; Artman et al 2002b; Baldwinn 2001; Sullivan 2002).

The fraction of heart malformations that support intrauterine circulation compromise the spectrum of congenital heart defect (CHD). The anatomic features of most CHD in humans have been carefully catalogued. Recent advances in genetics and molecular biology stimulated a renaissance in studies of cardiac development. Genetic alterations and null mutations have targeted the heart and vascular system and established abnormalities in cardiovascular ontogeny as a primary cause of embryonic demise. The next era of pediatric cardiology will merge the genetic basis of cardiac development with directed therapy and prevention (Sullivan 2002; Wren et al 1999).

The incidence of congenital heart disease is generally stated to be about 8 per 1000 births. However, most report worldwide have not included individuals whose lesions were not recognized by clinical examination alone. In recent years it has become evident, based on ultrasound studies, that the incidence is considerably higher than that usually quoted. Prenatal ultrasound examination has documented not only congenital heart lesions, but also cardiac arrhythmias and myocardial dysfunction as important causes of fetal morbidity and mortality. In fact, cardiac anomalies and arrhythmias are the dominant cause of non-immune hydrops fetalis. Although there is such a high prevalence of heart disease, it was recognized, prior to the modern era of early evaluation and treatment, that the major mortality occurred during infancy. Thus, of all deaths related to congenital heart disease, 50 percent occurred by six months, and 80 percent by 1 year of age. It is thus appropriate that attention should be focused on heart disease in the neonate (Friedman and Silverman 2001; DeKeyser 2004; Ontoseno 1996; Wren et al 1996).

HISTORY

The newborn with cyanotic congenital heart disease typically has a benign birth history. Either congenital cardiovascular disease, pulmonary hypertension of the newborn, or severe intra or extraparenchymal lung disease cause cyanosis. The primary differential diagnosis of congenital heart disease is persistent pulmonary hypertension of the newborn. The cyanotic infant with primary parenchyma lung disease usually has severe respiratory distress requiring mechanical ventilation and has an abnormal chest radiograph. The infant with pulmonary hypertension may only have mild or moderate respiratory distress, and a perinatal history of birth asphyxia, with or without meconium aspiration. Additionally, the infant may be small for gestational age, or the mother may have taken non-steroidal anti-inflammatory medications over the weeks prior to birth, which can cause constriction of the duct's arteriosus and subsequent pulmonary hypertension (Artman et al 2001b; Korones and Bada-Ellzey 1993; Rebecca et al 2003).

The ductus arteriosus usually maintains adequate blood flow and mixing immediately after birth, the Apgar scores are normal (or nearly so), and cyanosis is not suspected. It is not until hours or days after birth that the newborn becomes cyanotic, frequently during feeding or while crying. The increased physical effort associated with feeding or crying increases oxygen consumption and decreases pulmonary blood flow, which causes cyanosis. Despite the presence of cyanosis, a history of respiratory distress is usually not obtained. The chemoreceptor response to hypoxemia is intact so that

mild tachypnea often occurs, but respiratory distress (e.g., retractions, nasal flaring, grunting) is usually not present because ventilation is normal (Artman et al 2001b; Korones and Bada-Ellzey 1993; Wilkinson 2002)

Table 1. Genes Implicated in the Etiology of Congenital Heart Defects (Srivastava and Baldwinn 2001; Sullivan 2002; Wren et al 1999)

Gene	Chromosomal Location	Function	Cardiac Defects
TBX1	22q11.2	Transcription factor	TOF, PTA, IAA
TBX5	12q24.1	Transcription factor	Conduction system defects, ASD, VSD, TOF,PTA, single ventricle
Nkx	2.5 5q34	Transcription factor	Conduction system defects, ASD, VSD, TOF, PTA, Epstein anomaly, HLHS, AS, CoA, heterotaxy
GATA	4 8p23.1-p22	Transcription factor	ASD
ZFPM2/FOG2	8q23	Transcription factor	TOF
JAG1	20p12	Cell signaling molecule	TOF, PS, PPS
PTPN11	12q24.1	Cell signaling molecule	PS, hypertrophic cardiomyopathy
LEFTYA	1q42	Cell signaling molecule	Heterotaxy
ACVR	3p22-p21.3	Cell signaling molecule	Heterotaxy
CF	C1 2	Cell signaling molecule	Heterotaxy, TGA
ZIC3	Xq26.2	Transcription factor	Heterotaxy
CRELD1	3p25-pter	Cell adhesion molecule	Heterotaxy, AVSD
Elastin	7q11.23	Structural protein	SVAS, PAS, TOF

Early discharge to home of newborn is increasingly common. It is thus important to recognize even mild cyanosis on the first or second day of life because progression to severe hypoxemia may not occur until after discharge. If there is any question, then pulse oxymetri should be performed to determine oxygen saturation. The transient desaturate must be distinguished from newborn with cyanotic heart disease who may also appear dusky initially only with crying or feeding (Sao Paulo 2001; Sastroasmoro 1989). Family history of congenital heart disease is relevant to the outcome of the fetus. Perhaps 50% of cardiac defects have a genetic origin. Thus, a positive family history increases the risk for subsequent children (Anderson et al 1987, Ontoseno 2003).

CLINICAL PRESENTATION OF CYANOTIC CONGENITAL HEART DISEASE
(Problems that cause too little blood to pass through the lungs)

These defects allow blood that has not been to the lungs to pick up oxygen (and, therefore, is oxygen-poor) to travel to the body. The body does not receive enough oxygen with these heart problems, and the baby will be cyanotic, or have a blue coloring.

Cyanosis is a critically important clinical finding in the newborn. It is the primary symptom of the most common forms of heart disease that present symptomatically in the newborn. If cyanosis due to congenital heart disease is not recognized the newborn may experience rapid and severe cardiovascular decompensation (Srivastava and Baldwinn 2001; Wilkinson 2002).

The critical features of cyanotic congenital heart disease are (Artman et al 2002b; Hsia 1998; Sastroasmoro 1989) : (1) systemic arterial hypoxemia is manifested clinically by central rather than peripheral cyanosis; (2) cyanosis is often not present immediately after birth, particularly in newborn who have defects that cause decreased pulmonary blood flow because the ductus arteriosus is still widely patent; (3) cyanosis is not evident until a significant amount of reduced hemoglobin is present. If the newborn has a systemic arterial oxygen saturation above 85%, cyanosis may be quite difficult to detect by visual inspection, Oxygen saturation should be measured by pulse oximetry if there is is any suggestion that even mild cyanosis may be present; (4) cyanosis is not evident until a significant amount of reduced hemoglobin is present. If the newborn has a systemic arterial oxygen saturation above 85%, cyanosis may be quite difficult to detect by visual

inspection. Oxygen saturation should be measured by pulse oxymetri if there is any suggestion that even mild cyanosis may be present; (5) the source of systemic arterial blood must be considered when evaluating the newborn for cyanosis. Different ventricles may perfuse the ascending and descending aorta if the ductus arteriosus is patent. Pulse oxymetri should be performed on the right hand, which, in a normal aortic arch receives blood from the ascending aorta, and on either foot, which receives blood from the descending aorta. Different conditions are associated with different relationships in oxygen saturation between the upper and lower body and defining the relationship my be very helpful in identifying the specific defect causing cyanosis. If pulse oxymetri shows a decreased saturation in the right hand, it is prudent to measure oxygen saturation in an ear or nose because the blood flow at these sites always arises from the ascending aorta; (6) newborn with cyanotic heart disease are hypoxemic and thus breathe rapidly. However, they rarely have respiratory distress (no retractions or nasal flaring) and arterial CO2 levels are usually decreased because of hyperventilation. Thus, these defects are rarely confused with primary lung disease.

CLINICAL PRESENTATION OF THE NEWBORN WITH EXCESSIVE PULMONARY BLOOD FLOW (Problems that cause too much blood to pass through the lungs) (Artman et al 2002a, Artman et al 2002b; DeKeyser 2004; Westmoreland 1999).

These defects allow oxygen-rich blood that should be traveling to the body to re-circulate through the lungs, causing increased pressure and stress in the lungs. The primary symptom in these newborn is tachypnea, often accompanied by mild respiratory distress and the mechanism for the tachypnea is not known. More likely, the increased production of interstitial fluid with the attendant increase in lymphatic flow is the primary cause of tachypnea in these newborn. As blood flow increases, lymphatic flow increases similarly. If precapillary pressures are increased as well, as occurs in defects associated with elevated pulmonary arterial pressure, lymphatic flow increases further. The fluid is predominantly peribronchial, which may impair bronchial function. Airway size may decrease and airway resistance may increase, further increasing the work of breathing. In addition, the newborn may be more likely to develop wheezing. Newborn with normal lung size and function and no abnormalities other than heart disease do not develop tachypnea and failure to thrive until pulmonary blood flow is very high (usually exceeding 2.5 times the normal flow). In contrast, newborn with intrinsic pulmonary disease who also have excessive pulmonary blood flow may develop tachypnea and heart failure when pulmonary blood flow is only modestly increased.

A common situation is the newborn prematurely who develops bronchopulmonary dysplasia. In this setting, the presence of even a minor cardiac structural defect (such as an atrial septal defect) that would not cause symptoms in a normal newborn might result in significant symptoms. Furthermore, newborn with underlying lung disease will be severely compromised if a cardiac defect that causes a marked increase in pulmonary blood flow (such as a large ventricular septal defect) is present.

CLINICAL PRESENTATION OF THE NEWBORN WITH DECREASED SYSTEMIC PERFUSION (problems that cause too little blood to travel to the body) (Artman et al 2002b; Sao Paulo 2001; Sullivan 2002; Westmorelans 1999).

These defects are a result of underdeveloped chambers of the heart or blockages in blood vessels that prevent the proper amount of blood from traveling to the body to meet its needs. Includes obstructive heart disease or myocardial dysfunction from sepsis, hematological abnormalities (anemia and polycythemia), or endocrine/metabolic disorders such as hypocalcaemia, hypoglycemia, and metabolic acidosis. Neonatal sepsis is common, especially in the setting of prolonged rupture of the membranes. Hematological abnormalities are associated with placenta abruption, twin-twin transfusion, placental insufficiency, post-term delivery, or small-for-gestational-age newborn.

Endocrine/metabolic disease may have a positive history. Newborn with obstructive heart disease rarely have a positive perinatal history. The newborn is stable during the first hours of life, but eventually develops poor feeding, pallor, diaphoresis, and tachypnea with respiratory distress. This may occur as late as 3 to 4 weeks after birth, so it is extremely important for every infant to be carefully assessed at the time of discharge and at subsequent visits during the first month of life. Subtle findings of irritability, pallor, or diaphoresis may reflect inadequate systemic perfusion.

PHYSICAL EXAMINATION

The physical examination should be performed systematically. Each step determines whether the newborn falls into a specific mode of presentation (cyanosis, decreased systemic perfusion, or excessive pulmonary blood flow), and once determined, whether the newborn falls into a specific homodynamic category. Ancillary tests assist in establishing specific diagnoses

and defining the most appropriate therapy for each newborn (Rebecca et al 2003; Sao Paulo 2001; Sastroasmoro 1989).

The general examination includes vital signs and observation of the unclothed and warm newborn. Heart rate, respiration rate, blood pressure, and oxygen saturations are considered in conjunction with respiratory status, perfusion, and color. Weight, length, and head circumference are measured and plotted on growth charts to aid in identifying failure to thrive. Any postnatal decrease in weight percentiles when compared to length and head circumference should raise the possibility of heart disease (Anderson et al 1987; Sao Paulo 2001).

The first sign to asses on general observation is central cyanosis. Peripheral cyanosis (acrocyanosis) is common in newborn, and reflects the normal unstable peripheral vasomotor tone. Central cyanosis, which is indicative of arterial oxygen desaturation, is the important sign to recognize. Thus, vascular beds with little vasoconstrictor tone such as the tongue, gums, and the buccal mucosa should be evaluate (not the hands, feet or perioral region). It is also important to evaluate the patient during conditions such as feeding or crying which are most likely to produce central cyanosis (Artman et al 2002b). Cyanosis is difficult to perceive until arterial oxygen saturation is less than about 85% and decreased hemoglobin concentration makes detecting cyanosis more difficult. Thus, oxygen saturation should be measured if there is any question of cyanosis (Artman et al 2002; Artman et al 2002b).

Measuring oxygen saturation simultaneously in the right hand and lower extremity by use of two pulse oxymeters is necessary to evaluate whether different ventricles perfuse the upper and lower bodies, at least partially. A difference in oxygen saturation of only 3% to 5% may be significant, but most oxymeters are only accurate to within ± 2% to 3%. For this reason, it may be helpful to reverse the probes to ensure that any difference (or absence of a difference) is real and not just related to inherent variations in the probes or oxymeters (Artman et al 2002a; Artman et al 2002b; Dinarrevic et al 2000).

Cyanosis may be appreciated by careful visual inspection; decreased systemic perfusion is identified by examination of the extremities; and tachypnea is noted by observing the respiratory rate and pattern. The presence of a congenital cardiovascular malformation (or less commonly, a cardiomyopathy) must be considered in the differential diagnosis of any infant with one or more of these findings. A cyanotic newborn likely has underlying heart disease but may have a primary pulmonary disorder. A newborn with decreased systemic perfusion may be septic whereas the newborn who is breathing rapidly may be clearing prenatal lung fluid or have other more serious forms of pulmonary disease. Cyanosis due to a congenital cardiovascular malformation is often present at birth, but decreased systemic perfusion and tachypnea due to a hemodynamically significant congenital heart defect may not develop for several days (or even a few weeks) (Anderson et al 1987; Artman et al 2002b).

The respiratory status should be carefully evaluated. Infants who have isolated cyanosis are usually tachypneic but do not have respiratory distress. In contrast, increased pulmonary venous pressure and pulmonary edema cause respiratory distress in addition to tachypnea. In that case, intercostals and/or sub costal retractions, nasal flaring, and grunting may be observed (Artman et al 2002b; Sastroasmoro 1989).

Tachypnea is often a subtle finding that develops over days or weeks, as pulmonary vascular resistance and hemoglobin concentration decline. Thus, tachypnea immediately after birth, in the absence of signs of cyanosis or decreased systemic perfusion, usually points to pulmonary disease rather than to heart disease. Parents rarely appreciate that an infants is breathing more rapidly than normal. Poor feeding with associated failure to thrive and diaphoresis is common; murmurs may be absent. Thus, an infant with unexplained faiure to thrive, particularly in association with tachypnea and diaphoresis, should be evaluated for possible congenital heart disease (Anderson et al 1987; Artman et al 2002b; DeKeyser 2004).

Signs of decreased systemic perfusion, including the temperature and color of the skin, blood pressure, peripheral pulses, and capillary refill, in each extremity should be assessed next. Lower extremity pulses are more easily palpated in the feet than in the inguinal area. Blood pressure should be measured in the upper and lower extremities, and the lower extremity blood pressure normally is slightly greater than the upper extremity blood pressure. Thus, the systolic pressure in the right arm and either leg should be measured simultaneously. If the pulses are decrease and no blood pressure differential is detected, the carotid arteries should be palpated. If they are increased, the newborn have coarctation or interrupted of the aorta and right subclavian artery arising anomalously from the descending aorta (DeKeyser 2004; Korones and Bada-Ellzey 1993).

Heart murmurs are common in many normal newborn and may be absent in many newborn with symptomatic cardiovascular disease. Thus, the mere presence of a murmur is of little value to the examination. However, specific murmur are much more likely to be appreciated

if the clinician has a differential in mind at the time that auscultation is performed. Moreover, the presence of a non-specific murmur is of much less concern in an infant who has an otherwise normal examination (Anderson et al 1987; DeKeyser 2004). The abdomen should be palpated because liver enlargement is often a sign of right atrial hypertension or increased circulating volume from excessive pulmonary blood flow. The location of the liver and stomach is reversed in situs inversus (DeKeyser 2004).

The cardiac examination begins with palpation of the precordium to evaluate the right ventricular pressure and volume load. The parasternal and subxiphoid impulses are increased in most newborn with cyanotic congenital heart disease. A palpable left ventricular impulse usually indicates increased volume load as the ventricular cavity dilates and extends anteriorly and laterally. In contrast, increased left ventricular pressure load often does not cause a palpable impulse (Artman et al 2002b; DeKeyser 2004). Unless an arrhythmia is present, an electrocardiogram is generally of limited value in making a specific diagnosis at birth.

The chest radiograph provides information about the heart size, pulmonary blood flow, and the lung parenchyma. Active pulmonary vascularity is often difficult to asses at birth because even when pulmonary blood flow is three to four times systemic flow, the pulmonary vessels may not appear large.

Echocardiography is the mainstay of the diagnosis of the newborn with symptomatic heart disease and has largely replaced cardiac catheterization. Is crucial to understanding each newborn with symptomatic heart disease. However, to understand the pathophysiology and often the best care, it is important to take a systematic approach at each level of the evaluation and use each piece of information to build upon that understanding (Anderson et al 1987; Artman et al 2002b; DeKeyser 2004).

INITIAL TREATMENT

(Artman et al 2002b; Lewis et al 1981; Rao 1998)

Timely initiation of medical therapy in newborn with critical congenital heart disease is necessary to prevent and/or reverse clinical deterioration. The general approach should follow the usual guidelines for management of a critical ill or potentially critical ill newborn.

Oxygen

Supplemental oxygen is often administered to newborn with known or suspected heart disease without full consideration of the goals of therapy and the possible adverse effect and must include setting endpoints of efficacy and toxicity.

Mechanical ventilation

Mechanical ventilation of the newborn whose primary symptom is cyanosis is often unnecessary. In contrast, mechanical ventilation and sedation are often very beneficial to the newborn with decreased systemic perfusion because decreasing or removing the work of breathing decrease oxygen consumption.

Fluid

Careful attention to fluid status and urine output is essential when managing newborns with critical congenital heart disease**.** In general, on the first day or two of life, newborn with congenital heart disease manifest the same fluid, glucose, and electrolyte requirements as infants without congenital heart disease. Depending on the particular defect, however, fluid and electrolyte management may change dramatically in the neonatal period. An infant with left-to-right shunting exhibit progressively greater left-to-right shunting as the pulmonary vascular resistance falls postnatally. Consequently, signs and symptoms of heart failure develop due to increase pulmonary blood flow, reduced systemic output, and compensatory sodium and water retention. Free water restriction and diuretic therapy are indicated to reduce total body sodium and water. Thus, it is important to constantly reassess and be willing to modify therapy as conditions change.

PROSTAGLANDIN E_1

(Dinarrevic et al 2000; Korones and Bada-Ellzey 1993; Lewis et al 1981; Mulyadi et al 2004)

The decision to initiate therapy with PGE_1 is usually not very difficult. Appropriate use of PGE1 is not only life-saving, but allows time for careful diagnosis, evaluation, and formulation of a rational treatment plan. The two general indications are either inadequate pulmonary blod flow because of pulmonary outflow obstruction (eg, critical pulmonary stenosis or pulmonary atresia). Or because of inadequate systemic blood flow caused by obstructed aortic flow (eg, critical aortic stenosis, coarctation or interrupted aortic arch, hypoplastic left-heart syndrome). In addition, PGE_1 is commonly used in infants with d-transposition of the great arteries to increased pulmonary blood flow. In this setting, the increased volume return to the left atrium promotes atrial left to right shunting, which will increase systemic oxygenation. If a newborn is suspected of having a structural defect for which either pulmonary or systemic blood flow depends upon flow through the ductus

arteriosus, then an infusion of PGE_1 should be started immediately. Newborn with ductus-dependent pulmonary blood flow generally present with severe hypoxemia. In contrast to newborn with lung disease, the arterial pO_2 will not increase significantly in response to administration of 100 % oxygen.

An infusion of PGE_1 should be started in any infant less than 2 weeks of age suspected of having cyanotic congenital heart disease. Newborn with ductus-dependent systemic blood flow typically present between 3 and 14 days of age and with signs of cardiogenic shock. In these conditions, dilatation of the ductus arteriosus allows the right ventricle to perfuse the ascending aorta.In general, any infant younger than 2 weeks presenting with shock, decreased pulses, cardiomegaly, and/or hepatomegaly should be considered a candidate for treatment with PGE_1. Confirmation of the diagnosis should not delay initiation of therapy. It is generally advisable to initiate PGE_1 therapy prior to transporting an newborn with suspected heart disease to a tertiary care center for more definitive diagnosis and treatment. If, after further evaluation, the newborn is found to not have structural heart disease, then the PGE_1 infusion can be discontinued.

Administration of PGE_1 will almost always maintain patency of the ductus arteriosus and will dilate a ductus that has recently constricted. However, PGE_1 will not open a ductus that is permanently closed and as such, will not be effective in older infants. Certainly, infant less than 2 weeks of age are candidates for treatment, but infant older than 4 weeks are much less likely to benefit. It is reasonable to attempt to open the ductus arteriosus in infant between 2 and 4 weeks, but the success rate is much lower than that in newborn. If the ductus has not reopened within 1 to 2 hours at a maximal dose of PGE_1 (0.10 µg/kg/min), then it is very unlikely that the ductus will open. The infusion should be discontinued and the infant should be considered for urgent surgical intervention.

HEMATOLOGIC CONSIDERATION

(Anderson et al 1987; Artman et al 2002b; Hsia 1998)

Newborn with congenital heart disease rarely have intrinsic hematologic problems. Cyanotic congenital heart disease may be associated with secondary hematologic abnormalities (e.g. thrombocytopenia), but these generally do not develop until later in life. However, it is important to note especially prone to iron deficiency. The hemoglobin and hematocrit alone may not sufficient for making this diagnosis, because a cyanotic newborn with “anemia” may actually have hemoglobin and hematocrit values within the ranges that are considered normal. It may be useful to measure the mean corpuscular volume (MCV) or serum ferritin levels. Typically polycythemic and they should not be allowed to develop iron deficiency. Iron deficiency, even without anemia, predisposes these newborn to thrombosis and cerebral vascular accidents.

PRINCIPLES OF MEDICAL MANAGEMENT

(Anderson et al 1987; Artman et al 2002b; DeKeyser 2004)

Providing medical care to a newborn with known or suspected heart disease can be overwhelming and anxiety provoking. Many practitioners tend to withdraw and to abdicate care to the pediatric cardiologist. This response is both unnecessary and counterproductive to the integrated team approach to optimal care. The care of the newborn with critical heart disease is no different from that newborn with other medical conditions in that application of a few general principles will promote effective care and minimize the chance of iatrogenic misadventures. It is imperative that a concerted team approach involving neonatology, cardiology, nursing, surgery, and anesthesiology is utilized. Effective and ongoing communication is essential for optimizing care and providing a uniform approach to the management of these complex medical patients.

TIMING OF SURGERY

(Artman et al 2002b; Mulyadi et al 2004; Rao 1998)

In general, cyanotic newborn can be established by administration of PGE_1 , and other supportive measure. Surgery should be scheduled on a semi-elective basis after careful evaluation is complete. One exception is noteworthy: surgery is the only effective therapy for neonates with obstructed total anomalous pulmonary venous return and should be performed as soon as the diagnosis is established.

Additionally, patients with d-transposition of the great arteries who have restrictive patent foramen ovale often need emergent balloon atrial septostomy because of profound hypoxemia. This can be performed at the bedside under echocardiographic guidance. After successful septostomy these newborn are usually quite stable and corrective surgery should be delayed until any end-organic damage resolves.

Newborn who present with congestive heart failure and shock because of left-heart obstructive lesions (e.g., interrupted aortic arch) also can be established by administration of PGE_1 and other supportive care. These newborn are likely to have sustained end-organ damage because of decreased perfusion. Deferring surgery to

allow recovery of, for example, renal and liver function, allows complete evaluation of the newborn and decreases surgical morbidity and risk.

TRANSPORT AND ARRIVAL FROM THE OPERATING ROOM (Rebecca et al 2003; Sao Paulo 2001)

The physician and nurses who will be responsible for care of the infant after surgery must understand the anatomic defect and familiar with the preoperative evaluation and course. Information must be provided regarding (a) intraoperative findings; (b) the exact procedure performed; (c) length of time on cardiopulmonary bypass, aortic cross-clamp time, level of hypothermia, and circulatory arrest time; (d) available postoperative haemodynamic and echocardiographic data especially regarding residual lesions; (e) complications including arrhythmias and bleeding; (f) location of catheters, tubes, and temporary pacing wires and (i) airway and ventilatory status.
A warmer, intravenous infusion and syringe pumps, suction, monitoring cables, and appropriate ventilator are set up in the intensive care unit before the patient's arrival.

NURSING CARE OF THE NEWBORN WITH CRITICAL HEART DISEASE (Anderson et al 1987; Dinarrevic et al 2000; Rebecca et al 2003)

The diagnosis of CHD evokes many emotions for parents, siblings, grandparents, and other family members. Nurses play an essential role in caring for the patient and in providing information to the family about the patient's condition and plan of care, coordinating support systems, and assisting parents in establishing and maintaining the parenting role.

Psychoneuroimmunology is the study of the interactions among behavior, neural, and endocrine functions and the immune system. It has been shown that the immune function of many patients in the intensive care unit is suppressed as a result of trauma, sepsis, or profound physiologic and psychological stress. Three of the most common stressors among patients with CHD in the intensive care unit are fever, hypoxia, and fear or anxiety. Findings have shown each of these stressors to be associated with decreased immune functioning. Nurses have an important responsibility to protect their patients from infection and promote their ability to heal. Several actions are suggested that can help the nurse achieve these goals. It is hoped that nurses would keep these interactions in mind while caring for their patients in the intensive care unit.

COUNSELING FAMILIES BASED ON ETIOLOGY AND EPIDEMIOLOGY OF CRITICAL HEART DISEASE IN THE NEWBORN (DeKeyser 2004; Ontoseno 1996; Rebecca et al 2003)

When a child is found to have congenital heart disease, the parents frequently have severe guilt feelings and are almost always worried about the risk of occurrence of congenital heart disease in future children. This approach must be correlated with all the other aspects of giving continued support with chronically ill children. Among the few environmental factors known to contribute to congenital heart defects are a virus and certain drugs. Women who contract rubella (German measles) during the first three months of pregnancy have a high risk of having a baby with a heart defect. Other viral infections also may contribute.

Certain medications also increase the risk. These include the acne medication Accutane, lithium (used to treat certain forms of mental illness) and, possibly, certain anti-seizure medications. Drinking alcohol in pregnancy also can increase the risk of heart defects babies with fetal alcohol syndrome (FAS) often have them. Studies also suggest that use of cocaine in pregnancy increases the risk of these birth defects.

Certain chronic illnesses in the mother also can increase the risk of heart defects. For example, women with diabetes are at increased risk of having a baby with a heart defect, although this risk can be reduced or eliminated if the diabetes is closely controlled, starting before pregnancy. Women with an inborn error of body chemistry called phenylketonuria (PKU) also are at high risk of having a baby with a heart defect, unless they follow a special diet before pregnancy and during the first trimester. Several studies suggest that women who do not consume enough of the B vitamin folic acid before and during the early weeks of pregnancy are at increased risk of having a baby with a heart defect.

It is necessary to counsel patients and families regarding recurrence risks, lifestyle, and long-term outcome for various forms of congenital heart disease. An understanding of the occurrence and etiology of CHD is essential to decreasing the disease burden and economic effects of this defects. Epidemiologic studies seek to measure disease frequency and to establish associations between disease states and multitude of other selected variables; for example, between coronary artery disease and serum cholesterol. These observational studies establish statistical associations (but not cause and effect) that are useful for (a) developing diagnostic screening studies; (b) defining heritability and recurrence risk; (c) developing testable hypotheses regarding etiology and pathogenesis; and (d) planning for effective delivery of healthcare services.

All epidemiologic studies begin with measures of disease frequency. The two most common measures are prevalence and incidence. Prevalence excludes those who have already died from CHD, those in whom the disease has been cured or in whom it has spontaneously resolved, and those with undetected disease. Incidence is expressed as a rate and is defined as the number of new cases among those at risk within a population over a certain period of time.

For CHD, the population at risk includes all embryos alive at the time the cardiovascular system is forming. Many heart defects are associated with spontaneous abortions and stillbirth. For example the prevalence of CHD is about 15% in fetuses that have been spontaneously aborted and about 8% in stillborn newborn. Thus, even with advances in fetal echocardiography, the true incidence of CHD is impossible to measure. The prevalence at live birth is often used as a proxy.

The method by which patients are identified affects the estimate of the number of cases. Suppose for example, that study A includes all patients referred to a tertiary care center, whereas study B includes all patients seen at age 5 years at a large health maintenance organization. The results of study A will necessary be biased toward more severe defects that cause symptoms. Milder forms of heart disease would possibly be missed and patients with less access to referrals may be excluded. The results of study B would not include those patients who had already died and those in whom a defect such as a ventricular septal defect had closed spontaneously. After a case of CHD is identified, the method of naming and classifying the defects will affect the results of epidemiologic studies. Unfortunately, no universally agreed-upon nomenclature and classification scheme exists. It is difficult, if not possible, to compare results from studies that have used different systems. Difficulties in calculating prevalence also result from determining the denominator or reference population. Characterizing an entire population at risk for developing CHD is quite difficult so a representative sample or subgroup is often selected.

GENETIC COUNSELING

(Anderson et al 1987; Rebecca et al 2003)

Question regarding causation are directly relevant to the risk of recurrence and the parents of a child with CHD will have questions about the risk of recurrence in subsequent pregnancies or in their grandchildren. Additionally, as more patients with congenital heart disease survive into adulthood, questions about recurrence come directly from patients. Unfortunately, completely accurate genetic counseling requires knowledge regarding causation and the direct cause of most CHD is not known. The availability of prenatal diagnosis including amniocentesis or chorionic villous sampling for chromosomal diagnosis and fetal echocardiography beginning at about 16 weeks gestation for evaluation of cardiac structure should be explained.

WHEN TO CALL THE DOCTOR ?

(Anderson et al 1987; Artman at al 2003b; Sao Paulo 2001)

Parents should not be expected to “diagnose” their baby’s problem per se, but should know to notify the physician if there is any change in their baby’s disposition, feeding, color, and so forth. It is rare to send a neonate home with a cardiac monitor. Documentation of the instructions and information that have been provided to parents at the time of discharge avoids confusion.

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