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PRINCIPLES OF MANAGEMENT OF THE NEONATE WITH CONGENITAL HEART DISEASE

By P. Syamasundar Rao, MD

INTRODUCTION

In a recent issue of Neonatology Today [1], an approach to the diagnosis of cyanotic neonate was presented. Once a cardiac baby is identified, a detailed echo-Doppler study should be performed and the diagnosis confirmed. Cardiac catheterization and selective cineangiography are rarely required for diagnostic purposes. In this review, principles of management of congenital heart disease (CHD) in the neonate will be discussed.

GENERAL MEASURES

During the process of identification, transfer to a tertiary care center and work-up, prevention of hypothermia, maintenance of neutral thermal environment, monitoring for and prompt treatment of hypoglycemia, treatment of hypocalcemia, monitoring acid-base status, treatment of metabolic acidosis with sodium bicarbonate (NaHCO_3), and management of respiratory acidosis with suction, intubation and assisted ventilation as deemed necessary are important and should be undertaken [1]. In patients with cyanotic CHD 30-40% O_2 is adequate and 100% O_2 is not necessary. If ductal dependant CHD is suspected, intravenous infusion of prostaglandin E_1 (PGE_1) should be started while waiting for confirmatory diagnosis.

Treatment of Cyanosis

No more than 30-40% humidified O_2 is necessary because of fixed intracardiac right-to-left shunting in cyanotic CHD. Metabolic acidosis ($\text{pH} < 7.25$), if present, should be treated. If there is marked hypercarbia ($\text{PaCO}_2 > 60$ torr) or respiratory depression, intubation and mechanical ventilation are indicated.

"Once a cardiac baby is identified, a detailed echo-Doppler study should be performed and the diagnosis confirmed. Cardiac catheterization and selective cineangiography are rarely required for diagnostic purposes."

In infants with severe right ventricular outflow tract obstruction, the pulmonary blood flow is ductal dependent. The ductus may be kept open by intravenous infusion PGE_1 .

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Table I. Ductal-dependent Cardiac Defects**A. Ductal-dependent Pulmonary Flow**

- Pulmonary atresia or critical stenosis with intact ventricular septum
- Pulmonary atresia with ventricular septal defect
- Severe tetralogy of Fallot
- Tricuspid atresia
- Complex cyanotic heart disease with pulmonary atresia or severe stenosis
- Ebstein's anomaly of the tricuspid valve
- Hypoplastic right ventricle

B. Ductal-dependent Systemic Flow

- Hypoplastic left heart syndrome
- Severe coarctation of the aorta syndrome
- Interrupted aortic arch

Various cardiac lesions which are dependent on patent ductus arteriosus (PDA) are listed in Table I. The current recommendations for infusion of PGE₁ are 0.05 to 0.1 mcg/kg/minute intravenously. While PGE₁ has been administered to infants from one day to 99 days, it is most likely to be effective the earlier in life it is begun. A small ductus may be made to dilate with PGE₁, but an already closed ductus may be difficult to reopen. Side effects include apnea, hyperthermia, muscular twitching and flushing. The side effects have not posed substantial management problems, but the neonate should be watched closely for apnea. We usually begin PGE₁ at the recommended dose of 0.05 to 0.1 mcg/kg/minute, but will rapidly reduce the dosage to 0.025 to 0.03 mcg/kg/minute once the oxygen saturations improve. This may avoid the need for endotracheal ventilation because of apnea.

If the cause of cyanosis is persistent fetal circulation, it should be managed accordingly. Additional treatment of cyanosis depends upon the specific cause (see below).

Treatment of Congestive Heart Failure

The treatment of congestive heart failure, including administration of inotropic agents, diuretics and after-load reducing agents is similar to that of older children [2] and will not be detailed here except to

state that the neonatal myocardial development is incomplete [3] and that the myocardial response to pre-load and afterload manipulations and inotropic agents is suboptimal.

Of particular importance is administration of PGE₁ to neonates with heart failure in conditions in which perfusion to lower part of the body (Table IB) is ductal dependent. The dosage and administration of PGE₁ are the same as described above.

SPECIFIC MEASURES

The measures undertaken will depend upon the specific physiologic and/or anatomic abnormality that the infant is identified to have. These are arbitrarily divided into physiologic and anatomic abnormalities, although there is considerable overlap.

Physiologic Abnormality

The type of therapy is dependent on the hemodynamic disturbance produced by the associated cardiac anomalies and may be discussed in terms of decreased pulmonary flow, increased pulmonary flow and intracardiac obstruction.

Decreased Pulmonary Blood Flow

If the pulmonary blood flow is decreased because of right ventricular outflow tract obstruction (Table IA), it may be aug-

mented initially with infusion of PGE₁ as detailed in the preceding sections. However, the effectiveness of keeping the ductus open with PGE₁ is temporary and the ductal sensitivity to PGE₁ decreases with increasing age. Therefore, a more permanent method of increasing pulmonary flow by surgery should be sought. Pulmonary blood flow may be increased by surgical creation of aortopulmonary shunts. Following the initial description of subclavian artery to ipsilateral pulmonary artery anastomosis by Blalock and Taussig in 1945 [4], a number of other procedures to increase pulmonary blood flow have been described and these include Potts shunt (descending aorta-to-left pulmonary artery anastomosis), Waterston-Cooley shunt (ascending aorta-to-right pulmonary artery anastomosis), central aortopulmonary fenestration or Gore-Tex shunt, modified Blalock-Taussig shunt (Gore-Tex interposition graft between the subclavian artery and the ipsilateral pulmonary artery), Glenn shunt (superior vena cava-to-right pulmonary artery anastomosis, end-to-end) and formalin infiltration of the wall of ductus arteriosus.

The classic Blalock-Taussig shunt [4] and its modification with an interposition Gore-Tex tube graft between the subclavian artery and the ipsilateral pulmonary artery, described by de Leval et al [5], have stood the test of time and are currently the procedures of choice for palliation of pulmonary oligemia in the neonate. Most surgeons prefer the modified Blalock-Taussig shunt.

In the patient with predominant obstruction at the pulmonary valve level (rare), balloon pulmonary valvuloplasty [6] may be used to increase the pulmonary blood flow. In patients with valvar pulmonary atresia perforation of the valve [7-10] followed by balloon dilatation may be attempted. Stenting the ductus arteriosus [10,11] is an attractive non-surgical option, but because of limited experience, it is not currently a first-line therapeutic option.

In summary, a number of palliative procedures are available to augment pulmonary blood flow in the neonate, but the modified Blalock-Taussig shunt is the recommended procedure of choice in most patients with complex CHD and severe right ventricular outflow tract obstruction resulting in pulmonary oligemia.

Table II. Complex Neonatal Heart Defects* Requiring Pulmonary Artery Banding

1. Swiss-cheese type of ventricular septal defects
2. Double-inlet left ventricle (single ventricle)
3. Double-outlet right ventricle
4. Tricuspid atresia with a large ventricular septal defect

* Without associate pulmonary stenosis

Table III. Interatrial Obstruction

1. Pulmonary atresia
2. Tricuspid atresia
3. Mitral atresia
4. Hypoplastic left heart syndrome
5. Total anomalous pulmonary venous connection

Table IV. Inter-ventricular Obstruction

1. Tricuspid atresia
2. Double-inlet left ventricle
3. Double-outlet right (or left) ventricle

Increased Pulmonary Blood Flow

Increased pulmonary blood flow and congestive heart failure can occur with simple ventricular septal defects (VSDs) (rare in the neonate), although it is more common with complex CHDs (Table II). Initially aggressive anti-congestive measures should be instituted. If there is ductal dependent systemic circulation (Table I B), PGE₁ infusion should be started as detailed in the preceding sections. Surgical constriction or banding of the pulmonary artery [12] has been used in the past in this subgroup of patients. But, because of advances in open heart surgery, including deep hypothermia, simple VSDs are repaired rather than banded. Banding of the pulmonary artery continues to be useful in the management of complicated heart defects without associated pulmonary stenosis, listed in Table II. Banding not only improves congestive heart failure but also helps achieve normal pulmonary artery pressure so that bi-directional Glenn and Fontan procedures can be safely performed later in the subgroup of patients who have single ventricle physiology. If associated aortic coarctation is present, the aortic obstruction must also be relieved.

Intracardiac Obstruction

This may occur at the level of patent foramen ovale (PFO) and VSD. Interatrial obstruction: In patients with atresia of the atrioventricular valves and/or atresia of semilunar valves and total anomalous pulmonary venous connection (Table III), the entire systemic or

pulmonary venous return, as the case may be, must pass through the PFO. Sometimes the PFO becomes obstructive. Balloon atrial septostomy [13] usually results in improvement. Blade atrial septostomy [14] is occasionally necessary, especially in older infants. More recently static dilatation of the PFO [15,16] has been used to achieve relief of interatrial obstruction. Surgical atrial septostomy is needed in some instances. If the atrial septum is intact (no PFO), perforation of the atrial septum either by Brockenbrough technique [17] or radiofrequency ablation [18] may become necessary; this is either followed by static dilatation [15,16] or placement of a stent [19,20].

Interventricular obstruction: Spontaneous closure of the VSD [21,22], causing interventricular obstruction can occur in certain complex cardiac defects (Table IV). If such VSD closure causes decreased pulmonary blood flow, the approach is similar to that described in Pulmonary Oligemia section (PGE₁ and modified Blalock-Taussig shunt). If the VSD closure causes obstruction to systemic flow, such obstructions should be relieved either by enlarging the VSD or the obstruction bypassed by anastomosis of the proximal stump of the divided pulmonary artery to the ascending aorta (Damus-Kaye-Stansel procedure [23]) directly or via a prosthetic conduct. Fortunately, development of interventricular obstruction severe enough to require interventions in the neonatal period is rare.

Anatomic Abnormality

Management of specific defects is largely based on anatomic abnormality; some defects can only be palliated as discussed in the preceding section and some defects can be and should be corrected either with transcatheter methodology or by surgical correction as deemed appropriate. These will be briefly discussed below. The order of listing and discussion is arbitrary and is neither related to importance nor frequency.

Critical Pulmonary Stenosis

The term critical pulmonary stenosis is applied when pulmonary valve obstruction results in supra-systemic right ventricular systolic pressure with resultant right to left shunt at the atrial level; these infants often have ductal dependent pulmonary circulation. Following initiation of PGE₁, percutaneous balloon pulmonary valvuloplasty should be undertaken [24-27]. If the procedure is not successful, surgical pulmonary valvotomy under direct vision should be performed.

Critical Aortic Stenosis

Very severe aortic valve stenosis with a large gradient, congestive heart failure or ductal dependent systemic circulation may be labeled as critical obstructions. Balloon aortic valvuloplasty is an acceptable alternative to surgery in the treatment of critical aortic stenosis in the neonate. Initially retrograde femoral arterial route was used for balloon aortic valvuloplasty [28-30]. Because of potential for injury of the femoral artery, alternative routes for accomplishing the procedure, namely, umbilical [31], carotid [32], subscapular [33] or axillary [34] artery and anterograde femoral venous [35,36] approaches have been attempted. More recently, anterograde, transumbilical venous route [37,38] has been introduced. Our preference is to use anterograde, transumbilical venous route initially and if that is not successful, retrograde, transumbilical arterial route is attempted, followed by carotid artery cut-down. In unlikely event of lack of success with these procedures, surgical valvotomy should be tried.

Coarctation of the Aorta

In coarctation patients medical management with anticongestive measures including PGE1 infusion to bypass the coarctation may be attempted. Despite immediate success following balloon angioplasty of aortic coarctation [39-41], the recurrence rate has been high. Because of the high recurrence rate and the association of aortic arch and isthmus hypoplasia, surgery by procedures such as extended isthmoplasty may be more appropriate. In special circumstances [42-44], when surgery is risky or contraindicated, balloon angioplasty may be attempted as an initial therapeutic option.

Infradiaphragmatic Total Anomalous Pulmonary Venous Connection

The majority of total anomalous pulmonary venous connection patients presenting in the neonatal period are of obstructive type, usually infra-diaphragmatic and require emergent surgical correction to include anastomosis of the common pulmonary vein with the left atrium. Non-obstructive types do not need correction as neonates.

Transposition of the Great Arteries

In transposition of the great arteries, the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. Consequently, these patients have parallel circulations instead of a normal in-series circulation. The fetal circulatory pathways (PFO and PDA) will initially provide some mixing, but they tend to undergo spontaneous closure, resulting in severe cyanosis. Initially, PGE1, and if necessary, Rashkind balloon atrial septostomy [13] may have to be performed to improve oxygen saturation. Balloon atrial septostomy followed by venous switch procedures (Senning or Mustard) between 3 to 6 months of age is now replaced by arterial switch (Jatene) procedure [45]. Balloon atrial septostomy at presentation, if necessary, followed by arterial switch procedure at one to two weeks of age is routine at most institutions.

Tetralogy of Fallot

While in the early years, palliation of pulmonary oligemia with Blalock-Taussig shunt followed later by total surgical correction between 2 to 5 years of age was advocated, the current approach is total surgical correction at presentation or when symptomatic unless there are anatomic or physiologic contraindications. If there is pulmonary atresia or other contraindications for total correction, then a modified Blalock-Taussig shunt [5] is performed.

Interrupted Aortic Arch

In patients with interrupted aortic arch, the initial management is PGE₁ administration, followed by either end-to-end anastomosis if the aortic arch can be mobilized or an interposition Gore-Tex graft to connect the both ends of aorta after resection of the ductal tissue. If a large VSD is present, it should also be corrected at the same time.

Ventricular Septal Defect

Initial medical management with anticongestive treatment followed by pulmonary artery banding is an approach of the past; now banding is replaced with total surgical correction. However, it should be noted that VSDs presenting with severe heart failure as a neonate is unlikely because of high pulmonary vascular resistance, although it can happen in the event of rapid drop in pulmonary vascular resistance.

Patent Ductus Arteriosus

This fetal circulatory pathway tends to close spontaneously after birth [3]. But in some infants, such spontaneous closure does not occur. This is more frequent in premature infants than in full-term babies. If the ductal shunt prevents normal recovery from respiratory distress syndrome, the PDA should be addressed. If fluid restriction and diuresis do not improve the respiratory status, prostaglandin synthetase inhibitors (indomethacin) should be administered to pharmacologically close the ductus. If the desired effect is not achieved, surgical ligation is indicated. Transcatheter methods are not useful in the premature infants. Pa-



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tients with ductal dependent congenital cardiac anomalies (Table I) do not need closure. The ductus should be kept open with PGE₁ infusion.

Patent Foramen Ovale

This fetal circulatory pathway also tends to close spontaneously after birth [3]. It may remain open in nearly 30% of normal individuals and does not need intervention. The role of PFO in other CHD is discussed in the respective sections.

Hypoplastic Left Heart Syndrome

Previously considered inoperable, there are two reasonable options at this time [46,47]: cardiac transplantation [48] and multistage Norwood correction [49]. Because of scarcity of cardiac donors, the majority of centers are performing stage I Norwood procedure initially followed by two-stage Fontan.

Stabilization with medical management including PGE₁ infusion and anti-congestive measures are instituted while decision on final treatment plans is instituted. The options of: Supportive care, Multistage Norwood procedure or Heart transplant are explained to parents in detail. Most parents appear to prefer multistage Norwood procedure. Administration of PGE₁ to maintain an open ductus, ensure adequacy of interatrial septal opening, no hyperventilation (maintain PaCO₂ at 40 mmHg torr), and no supplemental oxygen is recommended. Careful balancing of systemic and pulmonary circulations to avoid systemic hypoperfusion is needed [46,47]. Most of the time ambient oxygen concentration less than room air (14 to 18%) to increase pulmonary vascular resistance is necessary to maintain good systemic perfusion while waiting for surgery.

Because of relatively high mortality associated with stage I Norwood operation, alternative procedures such as Sano modification [50] and hybrid procedures [51,52] (stenting the ductus, banding the branch pulmonary arteries and opening the atrial septum by stenting it) are under active investigation.

Double-Inlet Left Ventricle and Other Single Ventricle Lesions

A large number of complex CHDs that have functionally one ventricle fall into this category and are candidates for Fontan [53] pathway. The Fontan operation can't be performed in the neonate because of high pulmonary artery pressure/resistance and, therefore, it becomes a multistage surgery

Initial palliation with PGE₁ infusion and modified Blalock-Taussig shunt in infants with pulmonary oligemia and pulmonary artery banding in patients with pulmonary plethora is undertaken. Bidirectional Glenn procedure at 3 to 6 months of age followed by Fontan conversion with an extra cardiac conduit at age 2 to 3 years is the current approach in their long term management.

Tricuspid Atresia

The approach to tricuspid atresia is similar to that used for the single ventricular lesions, just described.

Pulmonary Atresia with Intact Ventricular Septum.

The treatment objective is to achieve a four-chamber, bi-ventricular, completely separated circulation [10,54,55]. In the presence of right ventricular dependent coronary circulation, severe right ventricular hypoplasia and/or infundibular atresia, this objective can't be achieved. Palliation with PGE₁ administration followed by a modified Blalock-Taussig shunt to provide pulmonary blood flow and atrial septostomy to decompress the right atrium should be considered. If the right ventricle is tripartite and is of reasonable size, radiofrequency perforation of the valve [7-10] followed by balloon dilatation may be attempted. If this is not successful, surgical valvotomy is required. Despite successful opening of the atresia, some neonates may remain ductal dependent for a few days to few weeks.

Truncus Arteriosus

Initial attempts to correct truncus arteriosus by a two-stage approach (pulmonary

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artery banding in infancy followed by complete correction in childhood) have now been replaced with complete correction within the first few weeks to months [56]. This appears feasible with relatively low mortality although the homograft conduit may have to be replaced because of growth of the child and calcific degeneration of the conduit. Most of the babies may not need correction in the neonatal period.

Ebstein's Anomaly of the Tricuspid Valve

Cyanotic neonates with Ebstein's anomaly of the tricuspid valve who are otherwise asymptomatic do not need any treatment unless there is marked hypoxemia [57,58]. As the pulmonary vascular resistance and pressures fall, the cyanosis will improve. If severe hypoxemia is present, especially in association with right ventricular outflow obstruction, PGE₁ infusion may be helpful. As the pulmonary resistance improves with time, the need for PGE₁ may be obviated. Severe tricuspid insufficiency may be problematic, but surgery in the neonatal period is poorly tolerated.

Syndrome of Absent Pulmonary Valves

Absent pulmonary valve syndrome is a rare CHD, most often consists of a large VSD, pulmonary valve ring hypoplasia, absent or rudimentary pulmonary valve leaflets and most importantly, massive (aneurismal) dilatation of the main and branch pulmonary arteries [59]. The latter produces varying degrees of tracheobronchial tree compression. When this condition presents in the neonatal period with tracheobronchial obstruction, the prognosis is poor. If there is no improvement with adequate ventilatory support, surgical correction with aneurismorrhaphy, closure of VSD and enlargement of pulmonary valve annulus should be performed. Insertion of homograft in the pulmonary position has been suggested, but may not be necessary in the neonatal period. [59].

CONCLUSIONS

Because of advances in the neonatal care, noninvasive diagnosis, understanding pathophysiology of CHD, anesthesia, medical and surgical therapy, most CHD presenting in the neonatal period can either be corrected or successfully palliated. Rapid identification, prompt transfer to a tertiary care center equipped to manage complex CHD, immediate and accurate diagnosis and urgent and appropriate therapy is mandatory. The neonatologist caring for the cyanotic newborn should coordinate the complex care with the pediatric cardiologist and cardiovascular surgeon. After administering the supportive care, the treatment required, by and large, is determined by the specific physiologic and/or anatomic diagnosis. Prostaglandin infusion and modified Blalock-Taussig shunt for the neonate with pulmonary oligemia, anticongestive measures and banding of the pulmonary artery for babies with pulmonary plethora and transcatheter or surgical relief of inter-atrial obstruction as needed should be provided. When feasible, transcatheter and/or surgical correction should be offered.

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