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Congenital Heart Disease: Cyanotic Lesions with Decreased Pulmonary Blood Flow

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Introduction

Patients with cyanotic congenital heart disease and decreased pulmonary blood flow may have a spectrum of abnormalities of the tricuspid valve, right ventricle, pulmonary valve, and pulmonary arteries. Treatment pathways are undertaken with the goals of eliminating cyanosis and optimizing right ventricular function, growth, and development. The impact of each intervention on tricuspid and pulmonary valve integrity, right ventricular systolic and diastolic function, and pulmonary artery anatomy must be carefully considered to achieve optimal outcomes.

Initial Evaluation and Stabilization of the Cyanotic Neonate

In cyanotic neonates, an initial evaluation is undertaken to determine whether the etiology of the cyanosis is cardiac or noncardiac in origin. The algorithm used at Children’s Hospital in Boston to triage cyanotic newborns is shown in Figure 9.1. A brief review of the maternal, family, and gestational histories and a directed physical examination may provide clues that favor either cardiac or pulmonary disease. A chest radiograph (CXR), electrocardiogram (ECG), and hyperoxia test should be obtained in all neonates with unexplained cyanosis. The CXR should be inspected for heart size, signs of parenchymal lung disease, increased or decreased pulmonary vascular markings, and sidedness of the aortic arch. For example, a very large heart and decreased pulmonary arterial markings on CXR suggests severe Ebstein’s anomaly of the tricuspid valve (Figure 9.2), whereas a normal heart size and decreased pulmonary blood flow suggest tetralogy of Fallot, pulmonary atresia with intact ventricular septum, or tricuspid atresia. Those with increased pulmonary vascular markings may have truncus arteriosus or total anomalous pulmonary venous return.

During the hyperoxia test (also called an oxygen challenge test), an arterial blood gas measurement is obtained from the right radial artery while the neonate is breathing room air, and a second blood gas value is obtained after the patient breathes for 10 min at 100% inspired oxygen. The PaO2 is often between 25 and 40 mm Hg on room air. In 100% FiO2, the PaO2 will usually rise to >80 mm Hg in patients with pulmonary disease (provided that significant pulmonary artery hypertension is not present) but remain unchanged or only increase slightly in most neonates with cyanotic heart disease. The PaCO2 is typically mildly decreased in newborns with cardiac disease and mildly elevated in those with pulmonary disease. Note that the hyperoxia test cannot be used in isolation to exclude critical congenital heart disease, as some neonates with left-sided obstructive lesions may have a PaO2 >60 mm Hg in any extremity or a PaO2 >150 mm Hg in the right arm.

If the aforementioned screening tests are suggestive of critical congenital heart disease, an echocardiogram should be obtained in a timely fashion. If pediatric cardiology consultation is readily available, and severe cyanosis (SaO2 <80%) and metabolic acidosis (pH <7.3) are not present, then the echocardiogram may be obtained prior to initiation of a prostaglandin E1 (PGE1) infusion. If the neonate has mild cyanosis (SaO2 >80%) and the echocardiogram reveals anatomy that does not likely require prompt surgical or transcatheter intervention (e.g., some neonates with tetralogy of Fallot), then observation without PGE1 is warranted. However, if pediatric cardiology consultation and echocardiography are not readily available, a prolonged transport is anticipated, or the neonate is profoundly cyanotic, then PGE1 should be administered without delay. If it is unclear from the initial postnatal echocardiogram whether early intervention is needed, then it is preferable to withhold PGE1, and monitor the neonate’s systemic oxygenation as the ductus arteriosus closes. This strategy avoids exposing some neonates to the side effects of PGE1 and also allows for timely identification of neonates with cyanotic congenital heart diseases who do not have ductal-dependent pulmonary blood flow and can have surgical or transcatheter intervention deferred.

Prostaglandin E1

Prostaglandin E1 has been used since the late 1970s to maintain ductal patency in infants with critical congenital heart disease.
Prostaglandin E₁ will maintain systemic blood flow in neonates with severe left ventricular outflow tract obstruction, maintain pulmonary blood flow in those with severe right ventricular outflow tract obstruction, and allow for mixing of systemic and pulmonary blood flow in the setting of parallel anatomic circulation (e.g., D-transposition of the great arteries). Prostaglandin E₁ allows adequate time for interhospital transport, detailed cardiac evaluation, treatment of noncardiac disorders, and semielective scheduling of most cardiac interventions. Neonates who present with shock or severe cyanosis can be given time for recovery of end-organ function before cardiac intervention. Prostaglandin E₁ may be administered through a central or peripheral venous line.

A PGE₁ dose of 0.05–0.1 μg/kg/min is used when the ductus arteriosus is severely constricted or functionally closed and severe cyanosis exists. Lower doses (0.01 μg/kg/min) will safely maintain ductal patency. The most common side effect of PGE₁ is apnea, which occurs in a minority of neonates receiving the drug [1]. The risk of apnea is not an absolute indication for endotracheal intubation and mechanical ventilation. Aminophylline may minimize the occurrence of apnea and need for endotracheal intubation in neonates receiving PGE₁ [2]. Other common side effects are listed in Table 9.1. Uncommonly, the initiation of PGE₁ may cause a significant clinical deterioration. In neonates with congenital absence of the ductus arteriosus (e.g., tetralogy of Fallot with absent pulmonary valve syndrome; some infants with pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries), PGE₁ may lower systemic vascular resistance, decrease pulmonary blood flow, and thus exacerbate cyanosis.

In contrast to neonates with ductal-dependent systemic blood flow, the ductus arteriosus in those with right ventricular outflow tract obstruction is often somewhat restrictive and follows a tortuous course from the aorta to the pulmonary artery. Diastolic runoff from the aorta is thus diminished, and the risk for necrotizing enterocolitis may be diminished. Thus, it is reasonable to introduce enteral feedings in stable neonates with ductal-dependent pulmonary blood flow who are receiving PGE₁, provided that a reasonable diastolic blood pressure (>30 mm Hg) is present.