Coarctation of the Aorta: A Case Presentation

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IN THE U.K. EACH YEAR, THERE ARE APPROXIMATELY 4,600 babies born with congenital heart disease, or 1 in every 145 births. Their prognosis varies in accordance with the complexity of the congenital defect; however, mortality rates from surgery performed on children with congenital heart disease have declined by more than 50 percent since the late 1970s.[1] Of those infants born with congenital cardiovascular defects, it has been reported that 5 percent have coarctation of the aorta (CoA).[2] In Scotland, 75 percent of those babies born with congenital heart defects are diagnosed in infancy (Table 1). In North America, approximately 35,000 babies are born with a heart defect each year, and of these, 8 to 11 percent have CoA.[3]

Coarctation is a constriction or narrowing of the aorta (Figure 1), most commonly found distal to the left subclavian artery, opposite the point of entry of the ductus arteriosus (juxtaductal).[4] CoA can, however, occur in other locations along the aorta. It usually occurs in the thoracic and rarely in the abdominal aorta. There are several classification systems for CoA. The most useful classifies the blood flow by dividing the condition into pre- and postductal.

- Preductal CoA is often associated with major abnormalities. The coarctation is located proximal to the ductus arteriosus.
- Postductal CoA. The coarctation is located distal to the ductus arteriosus.

ABSTRACT

Coarctation is a constriction or narrowing of the aorta and presents most commonly within the first two weeks of life. This article reviews a case study of an infant diagnosed with coarctation of the aorta on day 8 of life. It includes an overview of the etiology, clinical presentation, and management plus an account of the infant's transport to a regional pediatric intensive care unit (PICU).

In infants with CoA and a coexisting patent ductus arteriosus (PDA), blood shunting usually occurs in two directions (bidirectional). Patency of the PDA may be critical for lower body perfusion in many infants.

A right-to-left shunt allows blood to flow from the right ventricle through the ductus to the aorta. A left-to-right shunt represents reverse flow from the aorta through the ductus to the pulmonary artery.

Neonates born with coarctation may be asymptomatic in the newborn period, with some remaining asymptomatic for years or decades.[5] The coarctation may be preductal or infantile, where the narrowing is proximal to the ductus arteriosus or ligamentum arteriosum and is more likely to be present in infancy. In postductal or adult coarctation, the narrowing is distal to the ductus arteriosus or ligamentum arteriosum and may be present in the adult. At all ages, prompt referral for specialist opinion and treatment greatly increases survival rates and decreases short-term and long-term health complications.[6]

CASE PRESENTATION

Baby A was a 3,180 g male infant born by spontaneous vertex delivery at 38 weeks gestational age. The mother, a 29-year-old gravida 2, para 1, had an uneventful antenatal period with normal prenatal ultrasound scans. She had normal serology and no history of sepsis. The labor and delivery were uncomplicated, and Baby A was born in good

TABLE 1 ■ Estimated Babies Born with Congenital Heart Disease, 2001, United Kingdom

Country	Total Births	Number of Babies Born with Congenital Heart Disease		
		Diagnosed in Infancy (5.2/1,000 Births)	Diagnosed 1–15 Years (1.7/1,000 Births)	Total (6.9/1,000 Births)
England and Wales	594,634	3,092	1,011	4,103
Scotland	52,527	273	89	362
Northern Ireland	21,962	114	38	152
Total	**669,123**	**3,479**	**1,138**	**4,617**

Note: Incidence rates, in parentheses, are from the Northern Region Paediatric Cardiology Database, 1985–1999.

From: British Heart Foundation. (2003). Congenital heart disease statistics. Retrieved June 2, 2007, from http://www.heartstats.org/uploads/documents%5CCoHDstatistics.pdf

condition, with no signs of respiratory compromise and Apgar scores of 9 at one minute and 9 at five minutes.

At his first newborn physical examination at 12 hours of age, Baby A appeared morphologically normal, with a normal cardiac examination. He bottle fed well over day 1 and was discharged home with his mother on day 2. Both mother and infant were reviewed daily by the community midwifery team. However, on day 8, the infant was noted by his mother to be increasingly tachypneic, and his feeding had deteriorated. The infant was evaluated by the general practitioner, who referred him to the regional NICU for ongoing care.

On clinical examination in the NICU, the infant was found to be irritable but alert. There was generalized cutis marmoratum and mild cyanosis in room air. He was tachypneic with a respiratory rate of 80 to 90 breaths per minute, a labored respiratory pattern, and mild subcostal retractions. Breath sounds were clear on auscultation. The heart rate was regular, and on auscultation, there was a grade 3/6 systolic murmur loudest over the left lower sternal margin, a gallop rhythm, and right ventricular heave. The femoral pulses were difficult to palpate, but brachial pulses were present. The liver edge was palpable 1 cm below the right costal margin.

At this point, a differential diagnosis was made on review of the clinical presentation and history. Mild cyanosis can result from cardiac, respiratory, or central nervous system disorders, other less common disorders, or sepsis.

Respiratory diseases to be excluded included lung disease such as pneumonia, air leak syndrome, and an undiagnosed congenital defect such as diaphragmatic hernia/abnormality. Cardiac diseases to be excluded were all cyanotic heart diseases and obstructive defects that can present with closure of the ductus arteriosus.

Cyanotic heart disease generally presents without respiratory symptoms, however, and can have effortless tachypnea.[7] Central nervous system diseases included periventricular–intraventricular hemmorhage, meningitis, and primary seizure disorder. Other conditions, such as sepsis/meningitis, hypothermia, and hypoglycemia had to be considered. However, the presence of a cardiac murmur and reduced femoral pulses indicated the high probability of a cardiac anomaly.

Further investigations included four-limb blood pressure, preductal and postductal oxygen saturation, and a hyperoxia test to assess cardiovascular symptoms. A chest x-ray was also ordered to review respiratory symptoms and assess heart size, shape, and vascularity.

A typical radiographic picture in cardiac disease would be an increase in the cardiothoracic ratio >0.6, suggestive of cardiomegaly. In preductal coarctation, generalized cardiomegaly with normal pulmonary vascularity is seen. With postductal CoA, an enlarged left ventricle and left atrium and a dilated ascending aorta are present.[7]

Chest x-ray reflected no air leak, generalized cardiomegaly, and a degree of pulmonary venous congestion, consistent with preductal CoA.

Blood was drawn for blood gas analysis, complete blood count, blood cultures, and chemistry studies. The infant was started on broad-spectrum antibiotics benzylpenicillin and gentamicin because sepsis was strongly suspected. An immediate cardiac referral was made in view of the clinical history, the absence of femoral pulses, and the audible murmur.

Results of the four-limb blood pressure reflected a gradient >15 percent, with the upper extremity pressures almost double those of the lower extremity pressures. The left arm mean pressure was 96 mmHg, left leg mean pressure was 47, right arm mean pressure was

FIGURE 1 ■ Coarctation of the aorta.

A, Postductal coarctation of the aorta. *B,* Diagrammatic representation of the common routes of collateral circulation that develop in association with postductal coarctation of the aorta. *C* and *D,* Preductal coarctation. *E,* Sketch of the aortic arch pattern in a 7-week embryo, showing the areas that normally involute. Note that the distal segment of the right dorsal aorta normally involutes as the right subclavian artery develops. *F,* Abnormal involution of a small distal segment of the left dorsal aorta. *G,* Later stage, showing the abnormally involuted segment appearing as a coarctation of the aorta. This moves to the region of the ductus arteriosus with the left subclavian artery. These drawings *(E to G)* illustrate one hypothesis about the embryological basis of coarctation of the aorta.
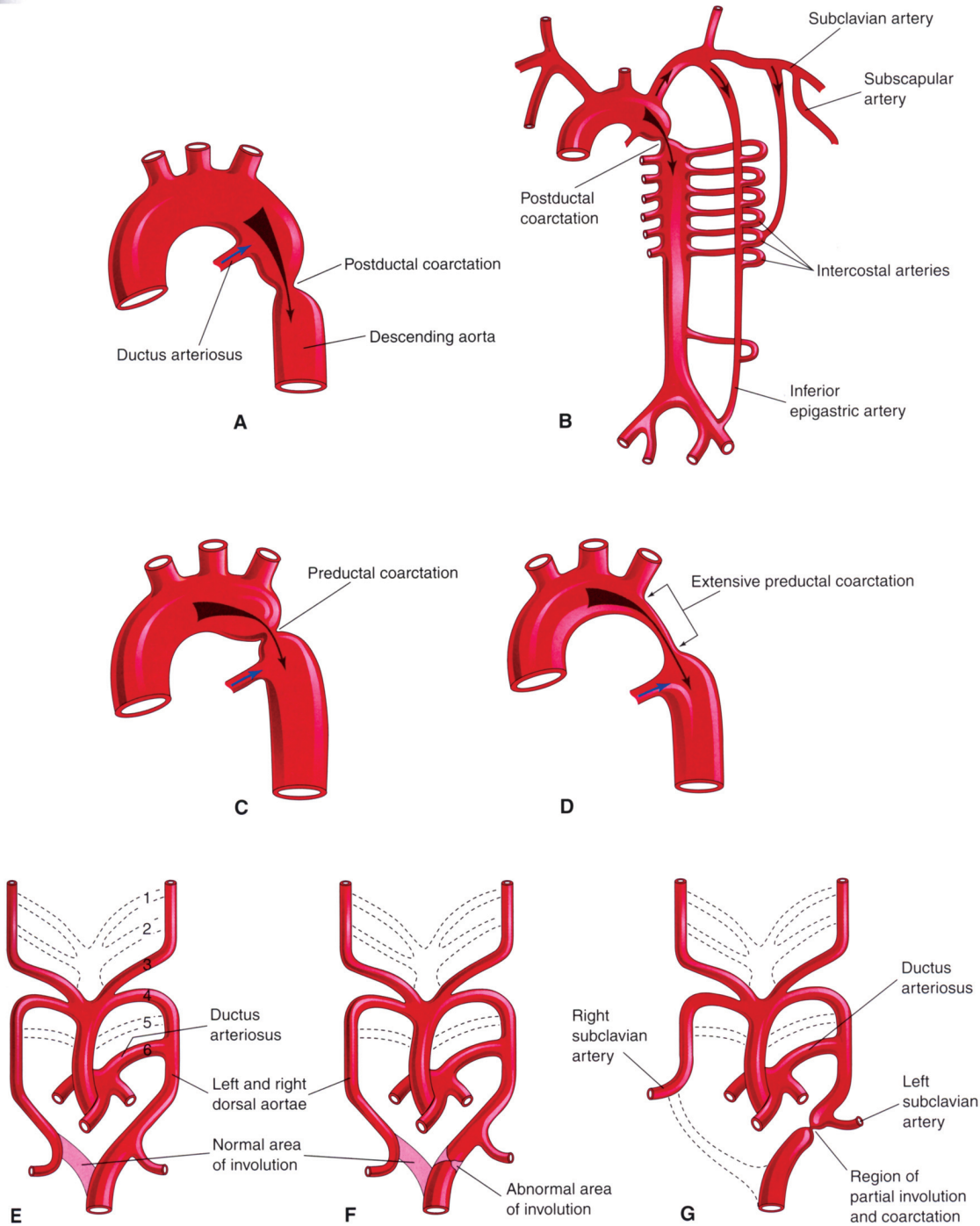
From: Moore, K. L., & Persaud, T. (2008). *The developing human–clinically oriented embryology,* (8th ed., p. 323). Philadelphia: Saunders. Reprinted by permission.

FIGURE 2 ■ Hemodynamics of aortic coarctation.
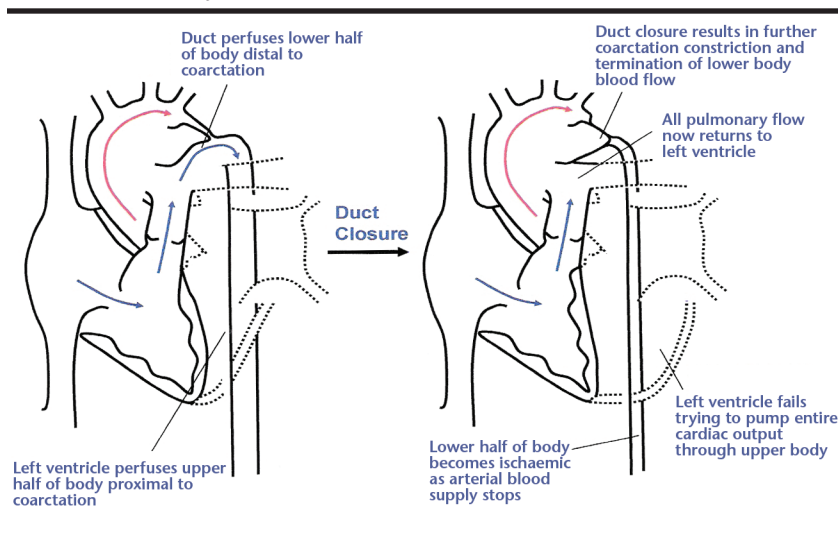
Duct = ductus arteriosus
Courtesy of Royal Hospital for Sick Children, NICU, Yorkhill, Glasgow.

90, and right leg mean pressure was 50. However, wide variations in blood pressure between limbs, which may be related to measurement techniques, such as using the wrong size blood pressure cuff, can present.[8] Capillary refill was reduced to three to five seconds, and the baby appeared poorly perfused. He was prescribed intravenous fluids of 10 percent dextrose at 135 mL/kg/day and given a bolus infusion of 0.9 percent normal saline at 10 mL/kg.

Initial arterial blood gas analysis demonstrated hypoxia with lactic acidosis, pH 7.29, partial pressure of carbon dioxide (PCO_2) 6.9 mmHg, partial pressure of oxygen 0.61 mmHg, base excess –5 mEq/liter (–5 mmol/liter), bicarbonate (HCO_3) 22.9 mEq/liter (22.9 mmol/liter), and elevated lactate 6.4 mEq/liter (6.4 mmol/liter). The hyperoxic test showed no significant difference in arterial oxygen levels in 100 percent oxygen, which was suggestive of congenital heart disease. Therapy was begun using a prostaglandin E_2 (PGE_2) infusion 20 ng/kg/minute to maintain patency of the ductus arteriosus. The infant was intubated after receiving morphine and suxamethonium (succinylcholine) and he was started on mechanical ventilation because of the presenting symptoms of respiratory compromise.

A diagnosis of coarctation of the aorta with a degree of aortic stenosis was made based on echocardiography. Cardiac contractility on echocardiography assessment was noted to be poor; therefore, a continuous infusion of dobutamine at a dose of 10 mcg/kg/minute was started to improve cardiac function. A request was then made for the neonatal transport team to transfer the baby to the regional cardiology center for ongoing management.

Baby A's general condition following intubation continued to deteriorate. A repeat arterial blood gas half an hour after intubation reflected a worsening metabolic acidosis: pH 7.16, PCO_2 5.5 mmHg, base excess –13.6, HCO_3 14.6 mEq/liter, and postductal oxygen saturation levels of 30 percent, preductal 55 percent. The prostaglandin infusion was increased to 100 ng/kg/minute in an attempt to open the ductus arteriosus and improve oxygenation prior to transfer. Laboratory blood test

results reflected elevated liver function tests—alanine aminotransferase 160 units/liter, aspartate aminotransferase level 70 units/liter—suggesting reduced lower body perfusion and evidence of hepatocellular damage. The serum sodium concentration was low at 123 mEq/liter (123 mmol/liter). Potassium was elevated at 30.5 mg/dL (7.8 mmol/liter), again suggesting compromised renal perfusion as a result of the coarctation.

Over the next 30 minutes, the postductal saturation level stabilized to around 75 percent, and Baby A was transferred to the cardiology center for review and ongoing management. He was transferred on mechanical ventilation and remained on 100 ng/kg/minute of PGE_2. The transfer was uneventful, and he was admitted to the PICU.

The infant was stabilized on prostaglandin therapy in the PICU. Two days later, he was taken to surgery, undergoing a successful end-to-end anastamosis. He had an uneventful postoperative course and was discharged home with follow-up at the cardiology clinic.

ETIOLOGY

Several theories have been generated to explain the development of aortic coarctation.[9–11] Two of the most widely accepted are the hemodynamic theory and the ductal tissue theory.[11]

The hemodynamic theory postulates that coarctation results from subnormal flow around the aortic arch. Lesions commonly associated with coarctation, such as ventricular septal defect, diminish the volume of left ventricular outflow in the fetus, decrease flow across the aortic isthmus, and promote the development of coarctation. Abnormal or obstructed embryonic blood flow through the left side of the heart can lead to underdevelopment of the aorta, valves, or chambers of the heart.[12]

The ductal tissue theory offers an alternative etiology. The ductus arteriosus diverts the circulation away from a large section of the aortic arch. Reduced blood flow with resultant vessel constriction forms an abnormal aortic segment consisting of thickened aortic media and intima known as the posterior shelf.[13] When the ductus arteriosus closes, ectopic ductal tissue pulls the shelf in toward the ductal orifice, causing narrowing.[14]

Malformed heart valves may also cause disturbances of flow. More than 60 percent of cases of coarctation have an associated bicuspid

aortic valve. Structurally abnormal valves and vessels frequently have a thickened intima, resulting in a turbulent blood flow and decreased circulation to distal vessel sites. This suboptimal circulation can lead to malformation and constriction.[15]

Several studies also imply an environmental influence in obstructions of the left ventricular outflow tract, with an association between chemical exposures, particularly to solvents, and coarctation of the aorta.[16,17] Wren and colleagues reported on temporal variability in birth prevalence of cardiovascular malformations, reflecting a trend toward a higher incidence of coarctation of the aorta over a 13-year period.[18]

Left heart lesions are frequently associated with coarctation; more complex structural cyanotic congenital heart conditions may also have an accompanying coarctation.[19] The term "complex coarctation" may be used to describe coarctation when associated with other cardiac lesions such as bicuspid aortic valve, ventricular septal defect, aortic stenosis, and secundum atrial septal defect.[20] Associated lesions have been reported in approximately 50 percent of patients diagnosed with CoA. CoA is more common in boys than girls in a ratio of around 2:1.[21] Coarctation may also be associated with Turner's syndrome and an increased incidence of cerebral artery aneurysm.[22]

Genetic factors are important in relation to patterns of familial recurrence of congenital heart disease.[23] Coarctation shows a weak familial association, with the recurrence risk in siblings being only 1:200.[24] This, however, may be an underestimate. A more recent review suggested that the recurrence was significantly higher if the affected parent was the mother.[25] A review of the Texas Birth Defect Registry found an increased prevalence of left ventricular outflow tract malformations and coarctation of the aorta in males that had a distinct racial/ethnic pattern not reported in previous studies. The authors reflected that all left ventricular outflow tract malformations demonstrated lower prevalence rate ratios for black males and hispanic males. However, increased CoA rates along the U.S.-Mexico border suggest environmental causes that will need more detailed study.[26]

PATHOPHYSIOLOGY

The branches of the aortic arch supply blood to the upper body (arms and head); those of the descending aorta supply the lower body (abdomen and legs). Coarctation is a narrowing of the aorta that is variable in its severity and location and becomes significant when there is a pressure gradient >20 mmHg across the aortic narrowing (see Figure 1).[27]

The narrowing or obstruction leads to an increased resistance to flow from the ascending aorta to the descending aorta with a subsequent drop in systemic blood pressure. The resultant hypoperfusion stimulates the kidneys to secrete renin, which constricts the arteries in an attempt to increase the distal pressure but can lead to hypertension in the proximal segment of the aorta. The lower limb perfusion is dependent on collateral circulation.[21]

Critical coarctation normally presents within the first few days of life if the lesion is severe and the ductus arteriosus closes. It can often be mistaken for septic shock due to the severity of symptoms and can be associated with a degree of isthmic hypoplasia.[28] The clinical presentation is frequently dependent on the amount of obstruction/degree of hypoplasia and the direction of the physiologic shunt.[15]

In a neonate, there is a great variability in the anatomy. A discrete narrowing may exist. Often a degree of arch hypoplasia is present, and the size and length of the aortic isthmus may vary. The origin of the left subclavian artery may also be involved in the narrowing, and an anomalous right subclavian artery rising beyond the coarctation may further complicate the situation.[20]

Closure of the ductus arteriosus in the newborn period usually occurs within the first two weeks of life. In coarctation of the aorta, patency of the ductus arteriosus is necessary to maintain systemic blood flow. The anatomic proximity of the open ductus to the coarctation widens the narrowed area, therefore decreasing obstructive resistance and increasing cardiac output. The severity of the coarctation is dependent on the length of the involved segment as well as the pressure gradient across the defect.[15] Ductal patency allows blood flow to bypass the narrow portion of the aorta, usually ensuring adequate flow to the descending aorta. Once the ductus closes, severe obstruction to flow beyond the left subclavian artery often results (Figure 2).

The severity of obstruction is directly related to the hemodynamics/systemic pressure gradient between the left ventricle and aorta in systole. In mild obstruction the gradient is <30 mmHg, in moderate, <50 mmHg, moderately severe 50–80 mmHg and in severe, >80mmHg up to a maximum of 200 mmHg. If there is an obstruction to the flow of blood out of the left ventricle, the pressure in the left ventricle will increase, leading to left ventricular hypertrophy.

Initially, when the ductus arteriosus is patent, the pressure in the pulmonary artery is equal to the systemic pressure, and the ductal shunt is consequently bidirectional. Where the narrowing is only mild, the pressure in the right ventricle is lower, the atrial shunt is left to right (aorta to pulmonary artery), and the saturations are the same on either side of the coarctation. Following ductal closure, the clinical consequences of the aortic obstruction become readily apparent. The left ventricle must then generate enough pressure to force 50 percent of the cardiac output past the constriction. This sudden increase in pressure may be poorly tolerated by the neonatal myocardium, resulting in an acute deterioration due to myocardial dysfunction and lower body hypoperfusion.[28]

In severe coarctation, the obstruction to blood flow through the aortic arch is so great that the predominant shunt across the patent ductus arteriosus is from right to left, with blood flow from the right ventricle bypassing the pulmonary artery, crossing the patent ductus arteriosus, and entering the descending aorta. This results in deoxygenated blood

TABLE 2 ■ Obstructed Left Heart Lesions Presenting in the Newborn Period

Condition	Cyanosis	Pulses	Auscultation	Precordium	Electrocardiogram	Associated Anomalies
Coarctation	None	Weak femorals	Gallop, murmur between scapulae, LVOT, systolic murmur	Active	RA+ RV+ T ↓ V5–6	Turner syndrome
Critical aortic stenosis	None	Weak	Gallop, EC, LVOT, systolic murmur	Active	LV+ ST ↓ T ↓ V5–6	Uncommon
Aortic arch interruption	None	Strong proximal to lesion	Gallop, LVOT, systolic murmur	Active	LV/RV+ T ↓ V5–6	Di George syndrome
Hypoplastic left heart	Mild	Weak femoral, stronger if DA open	Gallop	Active	Small LV voltages	Uncommon

Key: DA = ductus arteriosus; EC = ejection click; LV = left ventricle; LVOT = left ventricular outflow tract; RA = right atrium; RV = right ventricle; T = T wave, ST = ST segment; V5–6 = chest leads 5–6

Adapted from: Archer, N. (2005). Cardiovascular disease. In J. M. Rennie (Ed.), *Roberton's textbook of neonatology* (4th ed., p. 648). London: Churchill Livingstone. Reprinted by permission.

perfusing the lower half of the body. This can be detected clinically by a decrease in oxygen saturations from arm to leg as measured by pulse oximetry.

Aortic valve abnormalities often accompany coarctation.[19] In less severe forms of coarctation, the murmur of the aortic valve anomaly may be the presenting feature, with an aortic ejection click at the apex of the heart, possibly with an aortic systolic murmur.[29]

There are complicating factors in infants with CoA that contribute to their variable presentations and response to therapy. Lesions associated with CoA include patent ductus arteriosus, atrial septal defect, ventricular septal defect, hypoplastic left heart syndrome, transposition of the great arteries, mitral valve anomalies, and aortic stenosis.[30]

Infants with coarctation may have a small or underdeveloped left ventricle with reduced pumping capacity. This significantly affects circulation to the areas supplied by the distal aorta, including lower extremities, gastrointestinal organs, and kidneys. The heart therefore begins to compensate for this, resulting in right ventricular hypertrophy. Hypertrophy increases the pumping force of the right ventricle, raising pulmonary pressure as the heart attempts to drive blood through the ductus arteriosus. In critical coarctation, this can lead to right and left heart failure, metabolic acidosis, and death.[31]

PRENATAL DIAGNOSIS

Fetal echocardiography is a well-established method of diagnosing a wide variety of congenital heart defects.[32] It may be carried out because the family has a history of congenital heart disease or because of abnormalities detected during a routine detailed fetal anomaly scan. Coarctation of the aorta is one of the most difficult conditions to diagnose during the fetal period, primarily because of the patency of the ductus arteriosus and the parallel circulation that predominates prior to birth. Fetal echocardiography may indicate coarctation of the aorta due to asymmetry of the great arteries, the ventricles, or both, with dominance of the right heart structures.[33] However, there are no quantitative fetal parameters that differentiate among fetuses who present with cardiac asymmetry, those who will develop or have CoA, and those who will be normal.[32]

Early diagnosis results in improved preoperative cardiovascular stability and a reduction in surgical delays.[34] However, until recently, there was little published evidence that mortality in infants with congenital heart disease is reduced by prenatal diagnosis. A small retrospective review reported that accurate diagnosis of coarctation can be made prenatally, which may potentially avoid death at home in undiagnosed cases and improve the infant's preoperative condition.[35] Franklin and associates reported that three infants in their study group who were undiagnosed prenatally died at home. All three had a normal neonatal examination and were clinically well at discharge. The authors highlighted the importance of identifying the coarctation prenatally so that medications to maintain ductal patency are initiated soon after birth. They noted that severe deterioration with collapse or death occurred in 10 of the 22 undiagnosed cases, but did not occur in any of the prenatally diagnosed group. The study highlighted that antenatal diagnosis does not detect all cases of coarctation, particularly if there is no ventricular disproportion. A few cases of coarctation may present with both normal fetal ventricular proportions and normal ratios of transverse arch to pulmonary artery; however, the authors stated that these are unlikely to be ductal dependent and therefore not prone to circulatory collapse.[35]

CLINICAL PRESENTATION AND DIAGNOSIS

Coarctation, like many forms of cardiac disease, can present in the clinically well newborn. Neonates with coarctation may have a normal newborn examination.[31] As the patent ductus arteriosus narrows and cardiac demand

TABLE 3 ■ Purpose of Different Ultrasound Modalities

Ultrasound Modality	Purpose
2D imaging	Function Anatomic detail Wall thickness, size of chambers
Doppler	Direction/velocity of flow, flow patterns, and turbulence
Pulsed wave	Sampling in a localized region Risk of inaccuracies with high velocities
Continuous wave	Not precisely localized Good for high velocities
Color flow	Immediate identification of site of abnormal flow pattern Identification of regurgitation and small shunts
M mode	Function and dimensions

Adapted from: Archer, N. (2005). Cardiovascular disease. In J. M. Rennie (Ed.), *Roberton's textbook of neonatology* (4th ed., p. 628). London: Churchill Livingstone. Reprinted by permission.

increases, the infant becomes symptomatic. Right-to-left ductal shunting results in deoxygenated blood mixing with blood that supplies the lower extremities; however, as the ductus arteriosus closes, total blood flow to the lower half of the body is severely compromised. As a result, the infant's feet and legs may appear pale or cool to the touch. Femoral, popliteal, and dorsalis pedis pulses are weak or absent. In contrast, carotid, brachial, and radial pulses may be strong and full. In CoA diagnosis, Taylor has stated that a disparity between upper and lower limb values is the most significant sign.[36] Crossland and coworkers have reported a wide variation in blood pressure between limbs with the measurement techniques currently used.[8]

Symptoms such as poor feeding, tachypnea, and cool lower extremities may not become apparent until after discharge, usually coincident with closure of the ductus arteriosus. The timing and type of presentation depend on the site of the obstruction and the degree of narrowing, as well as the timing of closure of the ductus arteriosus.

Clinical presentation in the neonatal period and beyond into infancy may include a murmur, disparity of pulse intensity from upper to lower body, or the absence of femoral pulses, tachypnea, and cyanosis.[36]

The symptoms associated with severe infant coarctation are very similar to those seen in severe sepsis and collapse due to a metabolic disorder.[6] The differential diagnosis for neonates presenting with signs and symptoms of CoA includes a variety of obstruction malformations of the left ventricular outflow tract. These are summarized in Table 2.

Cardiac murmurs may be absent in up to half of infants with coarctation of the aorta.[31] A harsh systolic ejection murmur may be heard over the left sternal border and in the back, particularly over the area of the coarcted segment. A continuous murmur may be audible over the back or in the interscapular areas caused by the blood flow through collateral channels.[37] This presentation may be found in babies, but it is more commonly found in older children. The optimum place to listen for a murmur is in the upper left sternal border. The murmur may continue into diastole due to the continuous turbulent blood flow through the coarctation. A systolic ejection click may also be heard over the mid to lower-left sternal border, where a bicuspid aortic valve is present.[38] The infant may be tachypneic, and the precordium may be visually active due to the increased work of the heart against the constricted aorta. Additional heart sounds may be present; these usually constitute a gallop rhythm in any infant with tachycardia.[31]

Following closure of the PDA, the infant presents with more dramatic signs of congestive heart failure and circulatory collapse. Infants are generally acidotic, tachypneic, pale and shut down, with cool peripheries. A murmur and gallop may be present, and the liver and spleen edges may be palpable below the rib margins.

Coarctation missed in childhood accounts for a large proportion of adult patients with congenital heart disease that usually consists of a discrete narrowing beyond the origin of the left subclavian artery.[39] Most adults present with hypertension (sometimes in pregnancy), but some have poor femoral pulses or a systolic murmur detected during routine medical care. As age progresses, there is a propensity to develop ascending aorta aneurysms and thinning and weakening of the aortic wall, which may occur in segments of the aorta close to the coarctation.[20]
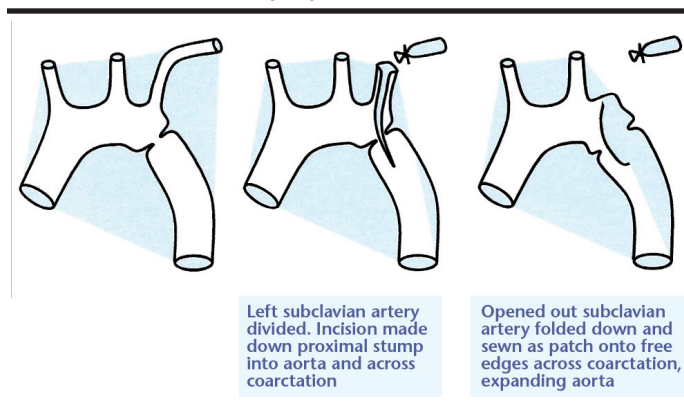
Echocardiography

Transthoracic echocardiography is the investigation of choice to confirm the diagnosis and identify the specific site of the coarctation. Different ultrasound modalities can be used to evaluate the cardiovascular system (Table 3). Anatomic detail is given by two-dimensional imaging. Color flow Doppler highlights areas of turbulent flow.

Pulse wave and continuous wave Doppler are used to determine the pressure gradient or the degree of narrowing.[15] The general function and physiologic condition of the heart are assessed by measuring the size and contractility of the main cardiac chambers and vessels.

Chest X-Ray

Chest x-ray may be nonspecific in relation to diagnosis in infants, often demonstrating cardiomegaly and signs of pulmonary congestion as seen in many forms of cardiac failure.[40] However, it can provide useful information that may contribute to differential diagnosis. The heart size can be reviewed by measuring the cardiothoracic diameter. A ratio of over 0.6 is indicative of cardiomegaly and may be evident in CoA. The heart chambers should be assessed to identify individual enlargement of each chamber if a lateral and anterior-posterior exposure is taken. Right ventricular hypertrophy may be evident in preductal CoA.

Left subclavian artery divided. Incision made down proximal stump into aorta and across coarctation

Opened out subclavian artery folded down and sewn as patch onto free edges across coarctation, expanding aorta

Courtesy of Royal Hospital for Sick Children, NICU, Yorkhill, Glasgow.

Enlarged great arteries can indicate problems such as pulmonary hypertension and aortic stenosis. Lung vascularity should be reviewed for increased pulmonary vascular markings found in conditions with excessive pulmonary flow such as ventricular septal defects or patent ductus arteriosus, which can occur with CoA.

Electrocardiogram

The electrocardiogram is often initially normal.[29] It shows rightward QRS axis as the right ventricle becomes hypertrophic and diffuse ST segment depression, indicative of increased afterload and left ventricular hypertrophy.

Oxygenation

In infants with right-to-left shunting across the ductus arteriosus, pulse oximetry values may reveal normal levels in the upper limbs and significantly lower levels in the feet while the ductus arteriosus is still open.[41] Preductal and postductal blood gas analysis can confirm this and exclude lower limb peripheral cyanosis as a cause of the saturation difference.

MANAGEMENT

Infants who are diagnosed and successfully treated before the onset of heart failure have increased survival rates, reduced recovery time, and fewer overall health problems.[13,15] In a retrospective review, Franklin and colleagues reported that both collapse and death were more common in postnatally diagnosed CoA. The authors concluded that antenatal diagnosis improved preoperative clinical condition and survival rate.[35]

Prenatal diagnosis facilitates optimum management at delivery. The infant may be delivered electively in a cardiology center where immediate expert assessment can take place. Treatment appropriate to the physiologic status of the infant can then be initiated in a timely manner. Antenatally diagnosed infants who are asymptomatic at delivery would be managed conservatively, and elective surgical intervention would take place after cardiology review.

The initial management of the infant with a probable diagnosis of critical coarctation is resuscitation, including intravenous fluid, intubation and ventilation, inotropic support, and prostaglandin E infusion. Positive end-expiratory pressure is useful in overcoming pulmonary venous desaturation as a result of pulmonary edema secondary to left atrial hypertension. In some critical coarctations, even high-dose prostaglandin E does not maintain ductal patency, and urgent surgical intervention is needed.[28] However, in less severely affected infants, a low-dose prostaglandin infusion and, if required, inotropic support may be all that is necessary until surgical intervention is carried out.

When aortic coarctation is diagnosed, maintaining blood flow through the ductus arteriosus is crucial to maintain oxygenation. The drug of choice in ductal-dependent cyanotic heart disease is prostaglandin (PGE_2) to keep the ductus arteriosus patent. Administration of prostaglandin should not be delayed if ductal-dependent cyanotic heart disease is suspected.[42]

Prostaglandin E Therapy

Prostaglandin E_1 and E_2 are potent vasodilators; PGE_1 is the licensed preparation, but an identical dose of PGE_2 is equally effective.[43] Prostaglandin is an eicosanoid molecule that acts as a local hormone in most tissues of the body. It works by binding to the receptors on target cell plasma membranes and stimulates or inhibits the synthesis of second messengers such as cyclic adenosine monophosphate. The effects of prostaglandins include altering smooth muscle contraction, blood flow, respiration, and nerve impulse transmission.[44] There is rapid deactivation of prostaglandin during passage through the lung; therefore, the half-life is less than one minute. Interruptions in prostaglandin E infusions due to problems such as mechanical obstruction can therefore cause a rapid deterioration in the infant's clinical condition.

Prostaglandin E when used at high doses may cause destabilizing apnea.[45] Therefore, it has been general practice to intubate newborns on prostaglandin E therapy for transport to regional units. However, a recent report has suggested that infants with suspected ductal-dependent lesions on low-dose prostaglandin may not require intubation for transport.[46]

Surgical Treatment

Surgical repair in symptomatic infants is usually carried out as soon as the infant is stabilized. But in a small number, medical treatment may be ineffective, and surgical repair must be undertaken in a critically ill, decompensated infant. Surgical options for repair of coarctation in the neonate include subclavian flap repair, excision with end-to-end or end-to-side anastomosis, and patch arterioplasty.

Subclavian Flap Aortoplasty. In subclavian flap aortoplasty, the aorta is exposed through a left lateral thoracotomy incision. The coarctation area is widened by an incision that

FIGURE 4 ■ End-to-end anastomosis of the aorta.

Please refer to the print edition of Neonatal Network

opens the aorta longitudinally across the coarctation. The left subclavian artery is then divided and the distal stump oversewn. The longitudinal aortic incision is continued up to the cut proximal end of the subclavian artery, which is then opened and folded down to form a patch over the coarctation (Figure 3). This results in widening of the area, facilitating normal blood flow through the aorta. An advantage of the subclavian flap procedure is the use of autologous tissue, resulting in the capacity for growth, a tension-free repair with limited dissection, and avoidance of circumferential scarring. A potential disadvantage of this procedure is the retention of ductal tissue, which may act as a substrate for renewed coarctation. Another disadvantage of this technique is that it does not resolve the problem of tubular hypoplasia, a zone of diffuse narrowing of the aortic arch. It has been suggested that if the coarctation is corrected, increased systemic flow may reverse the hypoplasia of the arch.[20]

Anastomosis. This procedure has the advantage of removing all ductal tissue.[20] It is also usually performed through a left lateral thoracotomy incision. The coarctation segment is resected circumferentially, and the proximal and distal ends of the aorta are anastomosed to each other (Figure 4). The proximal incision may be extended on the underside of the aorta to manage areas of proximal hypoplasia, in which case the distal end is anastomosed to the underside of the transverse arch in the end-to-side manner. A potential disadvantage of this procedure is the presence of a circumferential scar and suture line tension, which may result in rupture. The latter can be resolved by mobilization of the descending aorta and the head and neck vessels and releasing them from their pericardial attachment.[20]

Patch Arterioplasty. In this procedure, the aortic obstruction is incised and removed, and a prosthetic patch is inserted. This provides a tension-free scar with limited dissection; however, ductal tissue is not removed, which can result in an increased risk of recoarctation.[47] A major concern is the possible increased risk of postoperative aneurysm formation associated with the excision of the coarctation shelf.[48] Consequently, the procedure is now rarely used.

COMPLICATIONS

All three techniques have a low rate of morbidity and mortality, but a rate of recoarctation that is highest in neonates and infants, with no single technique being clearly superior.[49] However, more recent reports reflect a reduced recoarctation rate, as low as 3 percent, with the introduction of modifications to treat simultaneously the hypoplastic transverse arch. These treatments include end-to-end anastomosis and extended aortic arch repair.[50] The subclavian flap technique can lead to loss of the pulses in the left arm, which are eventually reconstituted via collaterals.[51] Long-term problems such as an increased risk of hypertension and difficulties with the aortic valve have also been noted.[3]

Nonsurgical Treatment

Balloon angioplasty is a nonsurgical intervention that is generally used for secondary repair following recurring coarctation. It is not routinely carried out due to the high incidence of recurrent obstruction, aneurysm formation, and iliofemoral artery injury.[52] If the repair is undertaken in the newborn period, there is a 10 to 20 percent chance of recoarctation.[28]

Transportation of the Cardiac Neonate

A major component of Baby A's care was transportation to a regional center. There has been little published on the specific issues involved in transport of newborn cardiac patients. Ductal-dependent cardiac lesions can present particular challenges in relation to PGE_1 treatment, destabilizing apnea, and mechanical ventilation.[46]

When specialized cardiology services are centralized to improve patient care, it is crucial that risks associated with transfer are addressed and that clinicians undertaking these transports are highly qualified and experienced in the transport of the acute neonate. Improved transport outcomes of cardiac patients have been reported in some centers since the use of intravenous PGE_1 increased.[53] A retrospective study reviewing the transport of cardiac infants stated that although major problems were not encountered during transport, there was still potential to optimize infants' condition on arrival and improve written documentation and clinical observation during transport.[54] In relation to air transport, one study reported a low transport-related mortality rate in its study group. A relatively poor outcome was reported in very low birth weight infants and infants with major congenital heart disease.[55]

Some centers practice telemedicine, in which two-way transmission of live video, still images, and echocardiograms allows a cardiologist at a remote location to view the baby, the electrocardiogram, chest x-ray, and a live

echocardiogram.[56] This development not only facilitates appropriate and timely transfer of neonates with congenital heart disease to regional specialist units, but also prevents unnecessary transfer of infants found not to have congenital heart disease. A further advantage is that with an accurate diagnosis the transfer team is able to optimize management of the specific lesion.

The West of Scotland Neonatal Transport Service transferred 90 neonates with suspected congenital heart disease between April 2004 and December 2006. Of this group, 36 required mechanical ventilation, and 40 were on prostaglandin treatment. Within the group requiring prostaglandin, 31 were ventilated, and 19 were on low-dose prostaglandin (≥20 ng/kg/minute). Coarctation of the aorta was diagnosed in 10 of the total transfers, with 5 being ventilated for transfer, 4 of whom were on prostaglandin. During this period, the transport service in conjunction with Regional Cardiology Services elected not to routinely ventilate neonates with suspected ductal-dependent congenital heart disease on low-dose prostaglandin. During this period, no adverse incidents as a result of this management were reported.[57]

Follow-Up Care

After surgical repair, most infants are discharged home within five days and follow a normal neonatal course. Routine follow-up visits are scheduled at the cardiology clinic. It is important to obtain a detailed history from parents in relation to the infants' respiratory effort, feeding pattern, any color changes when feeding or crying, and responsiveness. Developmental milestones can be assessed through observation of motor, sensory, and social skills. The physical examination should monitor growth through weight gain, length, and head circumference. Recoarctation may be indicated by reduction in femoral pulse volume, hypertension, and changes in heart sounds, respiratory effort, skin color, and perfusion.[36]

CONCLUSION

Coarctation of the aorta presents most commonly within the first two weeks of life. The ductal-dependent nature of the lesion means that acute circulatory collapse is a distinct possibility in this group of patients. It is therefore crucial that the clinical manifestations of CoA are recognized and the infant referred to the appropriate cardiology specialist as soon as possible. The benefits of such diagnostic techniques as four-limb blood pressure and the hyperoxia test are debatable and dependent on the preferences of individual cardiologists. However, these techniques may be useful in centers where echocardiography cannot be readily done. Undoubtedly, early diagnosis and prompt referal to specialist centers for treatment leads to increased survival rates and decreased short- and long-term complications. ✪

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