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REFERENCES
8 Wright RA, Yang F, Moore SW: Tuberculous infection in a vascular prosthesis. Arch Surg 112:79-81, 1977

Prostaglandin E1 Infusion in the Hypoplastic Left Heart Syndrome*

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Aortic infusion of prostaglandin E1 in a moribund infant resulted in marked clinical improvement. Appropriate studies were conducted and the diagnosis of hypoplastic left heart syndrome was made. Although the outcome was unaltered, the improved clinical condition allowed us to exclude potentially curable defects such as aortic coarctation and interruption.

Infusions of prostaglandins E1 (PGE1) and E2 have been used to maintain postnatal patency of the ductus arteriosus in infants with severe right ventricular outflow obstruction.1-8 By maintaining ductal patency, adequate pulmonary blood flow and systemic oxygenation can be achieved, providing the opportunity for more definitive treatment. More recently, prostaglandins have been used successfully in patients with severe aortic obstruction in whom ductal patency was required to maintain systemic bloodflow.6-9 Its use in the hypoplastic left heart syndrome, however, has not been reported specifically.

We describe the use of PGE1 in a moribund infant with the hypoplastic left heart syndrome. Although this entity is universally fatal, the transient palliation and improvement which resulted allowed us to exclude potentially correctable defects.

CASE REPORT

A 3,560 gram boy was born at an outlying hospital following an uneventful 40-week gestation. Apgar scores were 8 and 9 at one and five minutes. Initial evaluation revealed acrocyanosis and a short, nonspecific heart murmur along the left sternal border. Over the first 24 hours there was progressive tachycardia and tachypnea and the appearance of generalized cyanosis. The infant was transferred to our hospital at 30 hours of age with marked vascular collapse and severe metabolic acidosis (pH 6.99) which was unresponsive to intravenously administered bicarbonate. There was no urine output in the 12 hours prior to admission.

Physical examination showed a moribund infant with mottled skin, poor capillary filling, absent peripheral pulses and gasping respiration. Heart rate was 150/minute. The heart tones were faint and no cardiac murmur was heard. Despite adequate arterial oxygenation on 100 percent O2 (Po2 89 mm Hg), the pH was 7.01 with a delta base of -27. It was our impression that the infant had a ducal dependent cardiac lesion. A PGE1 (U10136, Upjohn) infusion at a rate of 0.1 µg/kg/min was immediately begun through an umbilical catheter with its tip positioned at the level of the ductus arteriosus. Within 15 minutes of beginning the infusion, femoral pulses were palpated, capillary filling was markedly improved and urine output commenced. The patient's color and level of activity improved markedly. Without the administration of additional bicarbonate, the arterial pH rose to 7.26.

With improvement in the clinical status, the diagnostic workup was completed. A chest roentgenogram showed cardiomegaly and pulmonary overcirculation. The electrocardiogram was normal for age. An echocardiogram revealed a small left ventricle, a single A-V valve and severe hypoplasia of the aortic root. The PGE1 infusion was temporarily discontinued so that a bedside aortogram could be performed through the infusing catheter. It revealed a hypoplastic ascending aorta and aortic arch (Fig 1). Contrast was seen in the aortic end of the ductus arteriosus, which was widely patent.

With the diagnosis of hypoplastic left heart syndrome

Figure 1. Aortogram performed by injecting 3.5 ml contrast medium (Renografin-76) through the umbilical arterial catheter reveals severe hypoplasia of the ascending aorta (arrows) and aortic arch. The widely patent aortic end of the ductus arteriosus is also visualized (arrow heads).
made, the condition was fully discussed with the parents and the FGE₁ infusion was permanently discontinued. The infant died shortly thereafter.

**DISCUSSION**

*In vitro*¹⁰,¹¹ and *in vivo*¹² animal studies showing the effectiveness of E prostaglandins in dilating the ductus arteriosus led to its clinical use in infants with ductal dependent, cyanotic congenital heart defects. Although most reported uses have been in infants with right ventricular outflow obstruction and diminished pulmonary blood flow,¹³-¹⁷ its usefulness in aortic obstruction has also been demonstrated.¹³-¹⁷

Patients with interrupted aortic arch, severe aortic coarctation or the hypoplastic left heart depend on right-to-left ductal shunting for part or all of their systemic blood flow. When ductal constriction occurs, as it almost always does, the diminished systemic perfusion rapidly leads to severe metabolic acidosis and generalized deterioration. Since aortic coarctation and interruption are amenable to surgical correction, the need for accurate diagnosis is obvious. Such infants, however, are frequently so ill by the time they reach a cardiac diagnostic center that clinical findings are not helpful in diagnosis and diagnostic procedures or surgical intervention are associated with a high mortality. By reestablishing and maintaining patency of the ductus arteriosus with a PG infusion, improved tissue perfusion and oxygenation result, permitting correction and reversal of the metabolic acidosis.

In the present case, the PG infusion resulted in prompt clinical improvement. The increased systemic perfusion, as evidenced by the appearance of femoral pulses and urine output and a rise in arterial pH, could only be attributed to reopening of the ductus arteriosus. Although this infant had a fatal and uncorrectable lesion, the ability to stabilize the child and carry out appropriate diagnostic procedures reduced the risk of missing a potentially treatable condition. PG infusion is certainly not indicated when the diagnosis of hypoplastic left heart syndrome is known. It can, however, provide the opportunity of diagnosing and treating other lesions that may be clinically indistinguishable in the critically ill infant.

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**REFERENCES**


**Echocardiographic Features of Mitral Obstruction due to Bacterial Endocarditis**

Mohsin Alam, M.D.; Joseph W. Lewis, M.D.; Sol D. Pickard, M.D.; and Sidney Goldstein, M.D.

We describe a patient with mitral valvular obstruction due to vegetative endocarditis. The diagnosis was made before surgery by M-mode and two-dimensional echocardiograms, which revealed a mass of echoes obstructing the mitral orifice. This was confirmed subsequently at surgery. Both modes of echocardiography are of value in the noninvasive diagnosis of mitral valvular obstruction due to vegetative endocarditis, a condition which may be amenable to surgery for valvular replacement.

Bacterial endocarditis involving the mitral valve often results in valvular insufficiency.¹³,¹⁴ Although significant mitral valvular obstruction resulting from vegetative endocarditis has been reported,¹⁵ the echocardiographic features of this entity have not been detailed. The purpose of this report is to describe a patient in whom mitral valvular obstruction caused by bacterial vegetation was diagnosed by M-mode and two-dimensional echocardiograms. The echocardiographic features of this entity will be discussed.

**Case Report**

A 65-year-old white woman with known insulin-dependent diabetes mellitus underwent left metatarsal amputation in October of 1976 for a gangrenous middle toe. The patient had undergone ten diastolic murmurs.

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