The designation "pulmonary atresia and ventricular septal defect" implies a biventricular heart, concordant atrioventricular connections, a large ventricular septal defect, a single outlet aorta, and no direct flow from the ventricular mass directly into the pulmonary arteries. The aorta may be more closely aligned to the left ventricle, to the right ventricle, or it may override the interventricular septum. A main pulmonary trunk may be present, terminating blindly at the ventricular mass. But how is blood conducted into the lungs? While the intracardiac anatomy and some aspects of the pulmonary circulation can be defined by cross-sectional echocardiographic examination, the challenge of these patients is to unravel the often complex nature of the pulmonary arterial supply. The consideration of how blood is conducted into the lungs of patients with pulmonary atresia is germane to many forms of congenitally malformed hearts including those with atrioventricular discordance and single-outlet aorta, complex cardiac malformations associated with right or left atrial isomerism, double-inlet ventricle; and hearts with absent right or left atrioventricular connection.

**Prevalence**

Data from the New England Regional Infant Cardiac Program provided a frequency of 0.042 per 1000 livebirths [41], while that of the more recently completed Baltimore-Washington Infant Study did not specifically mention pulmonary atresia with ventricular septal defect [29].

**Morphogenesis**

Kutsche and Van Mierop [71] studied the morphology of hearts exhibiting pulmonary atresia with and without ventricular septal defect. Addressing the morphology of the pulmonary valve, the diameter of the main pulmonary trunk, and the morphology and topography of the ductus arteriosus, they concluded that hearts exhibiting pulmonary atresia and ventricular septal defect reflect a maturational arrest that is considerably earlier than that of pulmonary atresia and intact ventricular septum. (See Chapter 19.) That specific embryologic factor involved in the etiology of pulmonary atresia and ventricular septal defect may be related to an abnormality of mesenchymal and/or neural crest migration.

There is an extensive literature defining the embryology of the pulmonary arteries. The main pulmonary trunk originates from truncal septation. That this is so can be gleaned from the "Rosetta Stones" of cardiac pathology: those hearts exhibiting normal truncal septation, but...
with right and left branch pulmonary arteries originating from the aorta [5a, 9, 14, 37]. In these hearts there is a main pulmonary trunk originating above a right ventricular infundibulum and connected to the descending aorta through a ductus arteriosus, but from the main pulmonary trunk there are no branch pulmonary arteries (Fig. 16.1). The pulmonary arteries in these extremely rare cases may originate from the ascending aorta, or from the descending thoracic aorta (representing persistence of primitive intersegmental arteries). The right and left pulmonary arteries are derived from proximal sixth arch derivatives and the ductus arteriosus from distal sixth arch derivatives. Vessels within the lung paren-