Cardiac Anomalies in the Fetus

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- Congenital heart disease • Fetal intervention
- Hypoplastic left heart syndrome • Pulmonary atresia
- Balloon dilatation • Valvuloplasty

As the most frequent congenital anomaly and the leading cause of death among infants in the United States, congenital heart disease (CHD) is an attractive target for fetal therapy. With the development of successful neonatal repair for many types of CHD over the last 20 years, earlier postnatal therapy to restore physiologic anatomy has been encouraged, and fetal therapy has become the next frontier. Concurrent advances in interventional catheterization and fetal imaging provided a foundation for the novel field of fetal cardiac intervention. This article focuses on the current status of in utero catheter interventions for CHD with particular interest in therapy for defects characterized by progressive stenosis or atresia of the semilunar valves, the aortic and pulmonary, with development of subsequent ventricular hypoplasia.

FETAL CIRCULATION

In the normal fetal circulation, oxygenated blood from the umbilical vein is able to stream efficiently through the foramen ovale (FO) to the left heart and up to the brain. The desaturated blood from the superior vena cava is directed through the ductus arteriosus (DA) back to the placenta for reoxygenation (**Fig. 1**). Because pulmonary

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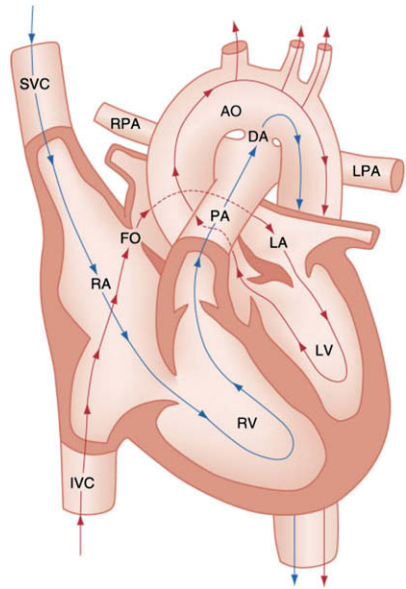

Fig. 1. Normal intracardiac fetal circulation. Physiologic shunting through the patent foramen ovale (FO) and the patent ductus arteriosus (DA). Oxygenated blood from the placenta (*red arrows*) reaches the right atrium (RA) by means of the inferior vena cava (IVC). This well-oxygenated blood is shunted preferentially from the RA across to the left atrium (LA) through the FO and then is ejected out the left ventricle (LV) to the ascending aorta (AO). Deoxygenated blood (*blue arrows*) returning from the superior vena cava (SVC) preferentially travels from RA into the RV, then out through the main pulmonary artery (PA). Because of the high pulmonary vascular resistance in the fetal lungs, this deoxygenated blood bypasses lungs and enters the descending aorta by means of the DA. (*From* Insaba AF. Cardiac disorders. In: Marx JA, editor. Rosen's emergency medicine: concepts and clinical practice. 6th edition. Philadelphia: Elsevier; 2006. p. 2568; with permission.)

vascular resistance is higher than systemic vascular resistance, blood shunts through the DA from the right side to the left, diverting around the lungs. At the moment of birth, however, pulmonary vascular resistance suddenly drops, the shunt reverses, and blood flows through the lungs. As the FO and DA close in the postnatal period, two distinct circuits are formed, the pulmonary and systemic vasculatures.

With severe semilunar valve pathology, however, there is only a single functional ventricular pump, the left ventricle in the case of pulmonary atresia (PA), and the right ventricle in the case of aortic stenosis (AS). In these situations, the DA is essential to continue perfusion to the systemic or pulmonary vasculature, and the FO is essential to allow the mixture of oxygenated and deoxygenated blood in the single ventricle system. As blood flows preferentially through the patent FO rather than the high-pressure ventricle, however, the reduction in flow through the ventricle retards growth, and in part, contributes to eventual hypoplasia.

In addition to semilunar pathology, if the atrial septum is intact (IAS) or severely stenosed, the circuit on the side of the atretic semilunar valve will become obstructed. In the extremely rare case of PA with a stenotic atrial septum, the fetal circulation is impaired and often not compatible with fetal life. In the case of established hypoplastic left heart syndrome (HLHS) with an intact atrial septum, the postnatal circulation is impaired and not compatible with neonatal life.