JACC Vol. 15, No. 3
March 1, 1990:678–81

Subcostal Two-Dimensional Echocardiographic Identification of Anomalous Attachment of Septum Primum in Patients With Left Atrioventricular Valve Underdevelopment

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Five variations of atrial septal morphology occur in hypoplastic left heart syndrome. One variety, termed anomalous attachment of septum primum, has been described only in necropsy series. Two-dimensional echocardiography was utilized to determine the incidence of this anomaly in patients with left atrioventricular (AV) valve underdevelopment, including those with other ventriculoarterial alignments, such as transposition of the great arteries.

Forty-eight (37%) of 129 patients with normally aligned great arteries (and two ventricles) had anomalous attachment of septum primum. Ten (34%) of 29 patients with double outlet right ventricle and left AV valve underdevelopment had this anomaly. Four (50%) of eight patients with single ventricle exhibited this atrial septal variant. The most reliable view to identify anomalous attachment was the subcostal left oblique-equivalent cut.

Recognition of atrial septal morphology has implications for preoperative and intraoperative management of patients with left AV valve underdevelopment. The similar prevalence of this atrial septal variant in patients with normally aligned great arteries, double outlet right ventricle and transposed great arteries suggests that there may be a common mechanism for left AV valve underdevelopment that is independent of the development of the arterial portion of the heart.

(J Am Coll Cardiol 1990;15:678–81)

Numerous technical refinements in cardiologic, surgical and intensive care management have improved early (1) and late (2) survival after palliative reconstructive surgery (the Norwood procedure) for hypoplastic left heart syndrome. One such development was the appreciation that there are five variations of atrial septal morphology (3) in this syndrome: congenitally small (or absent) foramen ovale, ostium primum atrial septal defect (atrioventricular [AV] septal defect), widely patent foramen ovale or true secundum type atrial septal defect, aneurysm of septum primum and anomalous attachment of septum primum. The recognition of these types is important in preoperative and intraoperative management.

One variety, termed anomalous attachment of septum primum (or, alternatively, posteriorly deviated superior attachment of septum primum), has been described only in necropsy series of hypoplastic left heart syndrome (3–6). This type of atrial septal morphology can be thought of as a malalignment-type atrial septal defect because the septum primum attaches directly to the posterosuperior left atrial wall, far to the left of the septum secundum.

We used two-dimensional echocardiography to determine 1) the incidence of this anomaly in patients with various degrees of left AV valve underdevelopment, and 2) the possibility that this atrial septal anomaly is not confined to patients with normally aligned great arteries but is also seen in other ventriculoarterial alignments (7), such as transposition of the great arteries.

Methods

Study patients. One hundred sixty-six consecutive infants <1 year of age with left AV valve hypoplasia, stenosis or atresia were examined with subcostal two-dimensional echocardiography before undergoing palliative reconstructive surgery. One hundred fifty-eight (95%) had two ventricles. Of these 158, 129 had normally aligned great arteries (7)

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Manuscript received July 26, 1989; revised manuscript received October 4, 1989, accepted October 10, 1989.

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0735-1097/90/$3.50

and 29 had double outlet right ventricle ({S,D,D} segmental anatomy, that is, situs *s*olitus of the atria, *D* ventricular loop, *D* position of the great arteries) with either subpulmonary infundibulum or bilateral infundibula (subpulmonary greater than subaortic) (8).

Eight patients (5%) had single (left) ventricle with outlet chamber (segmental anatomy was {S,L,L} in six, {S,D,D} in one and {S,D,S} situs *s*olitus of the atria, *D* loop of the ventricles, *s*olitus position of the great arteries in one).

Echocardiographic technique. A Hewlett-Packard 77020AC phased array system and 5.0 MHz short and medium focus transducers were employed. All patients were examined with three subcostal sweeps, as described previously (9,10).

Results

Normally aligned great arteries and two ventricles. Of the 129 patients with two ventricles and normally aligned great arteries, 48 (37%) had anomalous attachment of septum primum (Fig. 1, bottom). In all 48, the posterosuperior septum primum attached to the superior wall of the left atrium >4 mm to the left of the origin of the septum secundum and the posteroinferior attachment was normal. This anomalous attachment produced a flange partially covering the orifice of the right superior pulmonary vein; however, in no case did the septum primum attach to the left of the site of right superior pulmonary vein entrance, which would have resulted in partial anomalous pulmonary venous connection. Sixty-three (49%) of the 129 patients had either a patent foramen ovale or a secundum atrial septal defect (Fig. 1, middle), 5 (4%) had a congenitally small (≤2 mm) or absent foramen ovale (Fig. 1, top), 12 (9%) had an ostium primum atrial septal defect (as part of a common AV canal defect) and 1 (1%) had an aneurysm of the septum primum.

Double outlet right ventricle. Of the 29 patients with double outlet right ventricle, 10 (34%) had anomalous attachment of the posterior aspect of the septum primum. In 9 of these 10 patients, the posterosuperior attachment was displaced to the left; in the remaining patient, this attachment was normal, but the posteroinferior attachment was far to the left of the origin of the septum secundum. The anomalous attachment was so far to the left that the right inferior pulmonary vein drained into the right-sided atrium. Thirteen (44%) of the 29 patients had a patent foramen ovale or a secundum atrial septal defect, 5 (17%) had an ostium primum atrial septal defect, 1 (3%) had an aneurysm of the septum primum and none had a congenitally small or absent foramen ovale.

Single ventricle. Of the eight patients with single ventricle and left AV valve underdevelopment (seven of whom had associated transposition of the great arteries), four (50%) demonstrated anomalous attachment of the septum primum; in all four cases the anomalous attachment involved the posterosuperior portion of the septum primum.

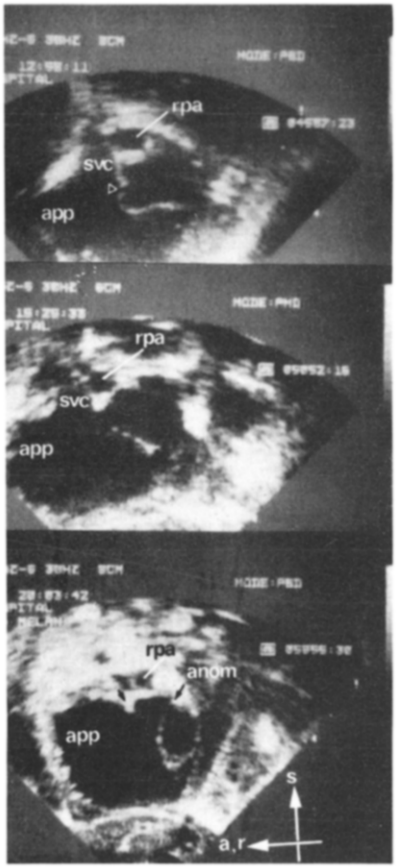

Figure 1. Subcostal left oblique views of three different atrial septal variations. **Top,** Congenitally small foramen ovale (**arrowhead**). **Middle,** Atrial septal defect of the ostium secundum type. **Bottom,** Anomalous (anom) attachment of septum primum far to the left of septum secundum (**unlabeled black arrowhead**). a,r = anterior and rightward; app = appendage; rpa = right pulmonary artery; s = superior; svc = superior vena cava.

Echocardiographic display. The most reliable echocardiographic view to identify anomalous attachment of the septum primum was the subcostal left oblique equivalent view (Fig. 1). Modified subcostal four chamber views not only failed to demonstrate the anomalous attachment, but also erroneously appeared to show septal dropout consistent with a secundum atrial septal defect (Fig. 2).

Discussion

Implications for preoperative management. Although ductus arteriosus narrowing or closure causes much of the metabolic derangement seen in the newborn with hypoplastic left heart syndrome, other structural and functional factors have an impact as well. One of the most important is the pulmonary vascular/systemic vascular resistance ratio.

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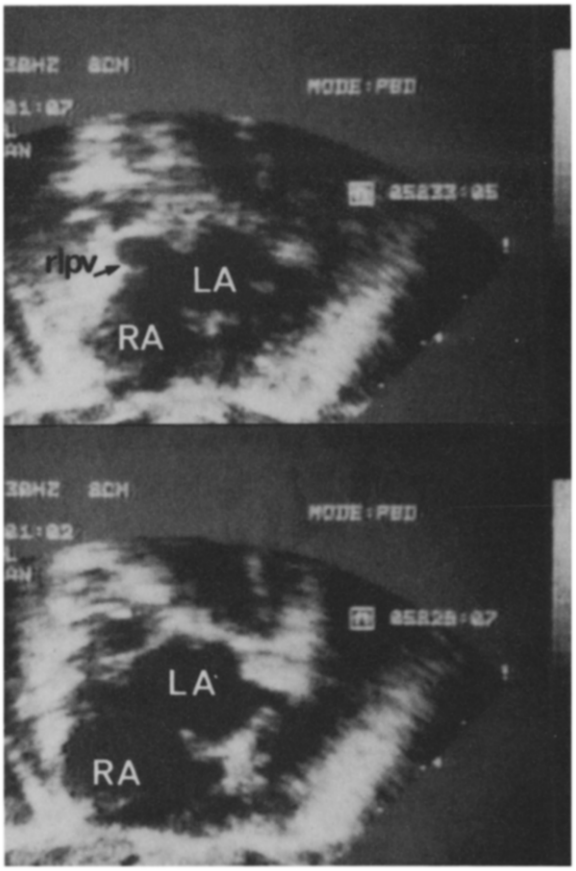

Figure 2. Subcostal frontal sweep. **Top,** At the most inferior portion of this sweep, the right lower pulmonary vein (rlpv) can be seen, along with what appears to be atrial septal dropout. **Bottom,** The transducer is angled more anteriorly; the right lower pulmonary vein is no longer visible, but the right upper pulmonary vein has not yet come into view. There now appears to be a large atrial septal dropout. These frames are from the same patient whose study is shown in Figure 1 (bottom), that is, with marked posterior and leftward displacement of the superior attachment of the septum primum. LA = left atrium; RA = right atrium.

The principal determinants of pulmonary vascular resistance are 1) the morphologic and functional characteristics of the pulmonary arterioles (for example, their responsiveness to changes in pH and partial pressure of carbon dioxide [P_{CO_2}] or to the administration of vasoactive drugs), and 2) the size of the atrial septal defect (or, in those with congenitally closed foramen ovale, the levoatrial cardinal vein).

An extremely restrictive atrial septal defect typically results in marked hypoxemia (for example, partial pressure of oxygen [P_{O_2}] in the 15 to 25 torr range). Two types of atrial septal morphology can result in such physiology in hypoplastic left heart syndrome: 1) congenitally small (or absent) foramen ovale, and 2) anomalous attachment of the septum primum. The former is associated with a thick muscularized atrial septum, and we have been uniformly unsuccessful at abolishing the left atrial-right atrial pressure gradient with various catheter techniques (11) in this group of patients.

Although the most superior aspect of the septum primum in patients with anomalous attachment is typically of normal thickness (3), the left atrial cavity size is much smaller than normal. Furthermore, the curvature of the septum primum tends to guide a catheter in the right atrium directly toward the posterosuperior left atrial wall, rather than toward the body of the left atrium. This theoretically could predispose to perforation of the left atrial roof. For these reasons, palliative surgery is performed emergently in newborns with an extremely restrictive atrial septal defect.

In the more usual case with a small to moderate size atrial septal defect, the pulmonary/systemic resistance ratio can usually be manipulated to yield a satisfactory P_{O_2} (35 to 45 torr) by the combination of avoidance of vasoactive drugs, hyperoxia and hyperventilation (12). Balloon atrial septotomy (11,13,14) is unnecessary, may be hazardous for the technical reasons outlined and, in some types of atrial septal morphology, may produce the undesirable result of a nonrestrictive atrial septal defect. (Although the latter would be a desirable result *after* the Norwood procedure, such a nonrestrictive defect makes preoperative management of the newborn with hypoplastic left heart syndrome difficult because pulmonary vascular resistance can decrease precipitously, resulting in pulmonary overcirculation.)

Implications for intraoperative and postoperative management. Resection of the atrial septum (1) at the time of palliation can be incomplete if this anomaly is not recognized; as a result, profound hypoxemia may occur early after operation. Incomplete initial resection may lead to a residual atrial septal gradient late postoperatively. Such a cause of pulmonary venous hypertension may also affect suitability for subsequent Fontan repair.

Echocardiographic methodology. Without use of the left oblique equivalent echocardiographic view, the diagnosis of anomalous attachment of the septum primum may be missed. The most likely reason for this phenomenon is the curvature of the atrial septum in cases of anomalous attachment (Fig. 3). In the sweep that includes the four chamber view and continues with progressive cranial angulation, the sector "transects" the atrial septum *only* during the initial portion of the sweep (Fig. 3, top, A). With progressive cranial angulation, the sector eventually contacts the atrial septum only tangentially (Fig. 3, top, B) and thereafter has no contact with the septum primum (Fig. 3, top, C). In a case without anomalous attachment (Fig. 3, bottom), the sector always transects the atrial septum until the superior vena caval-right atrial junction is displayed.

Embryologic implications. Finally, the similar prevalence of this anomalous attachment in patients with normally aligned great arteries, double outlet right ventricle and transposed great arteries suggests that there may be a

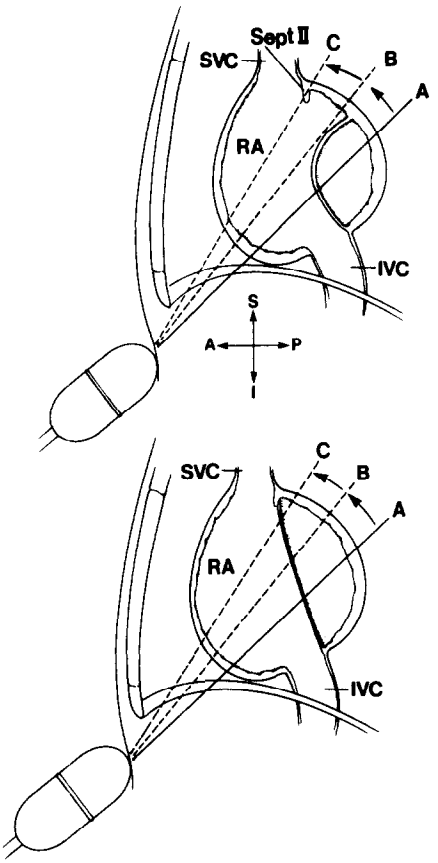

Figure 3. Lateral view of three different points (A, B and C) along the subcostal sweep that includes the four chamber view and continues with progressive cranial angulation. **Top,** Anomalous attachment of septum primum. The ultrasound sector crosses the atrial septum only during the initial portion of the sweep (A). By the time point B is reached, the sector no longer contacts any atrial septum; thus, the false appearance of an atrial septal defect occurs. **Bottom,** Normal attachment of septum primum. The ultrasound sector crosses atrial septal tissue (A, B) until the cranial angulation of the transducer continues *past* point C. IVC = inferior vena cava; Sept II = septum secundum; SVC = superior vena cava; other abbreviations as in Figure 2.

common mechanism for left AV valve underdevelopment that is *independent* of the development of the arterial portion of the heart.

We thank Beth Ann Aglira-Andrews, Audrey Alston-Jones, Jane Vetter and Mary Lou Weiss for superb technical assistance, and Kimberly Persick for secretarial assistance.

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