Left Ventricular Cine-angiocardiology in Endocardial Cushion Defect*

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Endocardial cushion defects are complex malformations with a wide clinical spectrum. The abnormalities range from a small defect of the atrial septum in the position of the ostium primum, with an abnormal cleft mitral valve, to more extensive abnormalities which include tricuspid incompetence and defects of the ventricular septum. In the most severe form a common atrioventricular canal occurs. The pathology determines the clinical and radiological picture (Campbell and Missen, 1957; Wakai and Edwards, 1956; DuShane *et al.*, 1960; Scott *et al.*, 1962).

In general, the defects can be divided into two clinical groups (Barnard and Schrire, 1961). Endocardial cushion defects, without a significant ventricular septal defect, consist of partial absence of the lower portion of the atrial septum and a cleft mitral valve. A small insignificant ventricular septal defect or cleft tricuspid valve may be present. In defects with a significant ventricular septal defect, the ventricular septal defect dominates the clinical picture, producing a large left-to-right shunt and often severe pulmonary arterial hypertension (Schrire *et al.*, 1963; Schrire and Vogelpoel, 1964). The electrocardiogram usually is diagnostic: it shows left axis deviation (-30° to -120°), a counterclockwise or figure-of-eight vector loop in the frontal plane, incomplete right bundle-branch block, and often a long P-R interval (Toscano-Barbosa, Brandenburg, and Burchell, 1956; Toscano-Barbosa, Brandenburg, and Swan, 1958).

Cardiac catheterization is mandatory, but, unless

it is undertaken with care, may not add much to the clinical findings. The presence of an endocardial cushion defect without a significant ventricular septal defect is confirmed by demonstrating a large left-to-right shunt at atrial level, normal or subnormal pulmonary vascular resistance, typical low passage of the catheter across the defect, and very similar drainage pathways of the pulmonary veins from each lung (Swan *et al.*, 1956; DuShane *et al.*, 1960; Braunwald, Lombardo, and Morrow, 1960). Left atrial cine-angiocardiology may add further information (Coelho, de Paiva, and Nunes, 1961; Gotsman, 1964; Astley, Gotsman, and Parsons, 1965). Defects with a significant ventricular septal defect have the haemodynamic pattern of a large combined ventricular and atrial septal defect, often with severe pulmonary arterial hypertension. Assessment of the degree of mitral and tricuspid incompetence may be difficult.

Recently the value of selective left ventriculography has been reported (Braunwald, Morrow, and Cooper, 1959; Baron *et al.*, 1964; Cornell, 1965; Omeri *et al.*, 1965; Girod *et al.*, 1965; Rubinstein *et al.*, 1966). We have undertaken left ventricular cine-angiocardiology routinely and have confirmed that certain features occur only in patients with an endocardial cushion defect.

This paper analyses the left ventricular cine-angiogram in 13 patients and relates the findings to the abnormal embryology and anatomy.

RESULTS

The ages of the 13 patients ranged from 6 months to 45 years and 12 were children under the age of 15. The diagnosis was confirmed at operation in 10. These 10 had a low defect of the atrial septum and a cleft mitral valve. Five also had a cleft tricuspid

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valve and 3 had a small ventricular septal defect—in one this was very small. Two had additional pulmonary stenosis. The other 3 patients did not need an operation: they had only a small atrial septal defect with an abnormal mitral valve. A correct clinical diagnosis was made before catheterization in each patient: the electrocardiogram was always diagnostic. Tricuspid incompetence was suspected in 3 of the 5 patients. Deficiency of the ventricular septum was also suspected in the 3 patients in whom it occurred.

The left-to-right shunt varied from 26 to 84 per cent. One patient with an insignificant ventricular septal defect had a 4 per cent rise in O_2 saturation in the right ventricle. The 2 patients with a significant ventricular septal defect had a 10 per cent rise in O_2 saturation in the right ventricle. The pulmonary vascular resistance ranged from 1–7 units. Two patients with significant defects in the ventricular septum had the highest vascular resistance (4 and 7 units, respectively). Left ventricular indicator dilution curves (sampling at a systemic artery) confirmed the presence of mitral incompetence in the 7 patients in whom this procedure was undertaken.

Abnormal Left Ventricular Cine-angiogram (Fig. 1 and 2). The deformed heart produced a characteristic appearance in the antero-posterior position. When the cardiac catheter was passed retrogradely from the aorta it was displaced to the left immediately below the aortic valve: the gentle curve concave to the left seen in the normal subject was altered, giving the catheter an elongated S-shaped profile.

The most important feature was an abnormal shape and configuration of the outflow tract. The normal left ventricle has a smooth ovoid contour, with the mitral and aortic valve orifices appearing as two chords of the ovoid. This shape was altered in every patient: the ventricle was divided into two halves—a smaller ovoid body and a narrow elongated outflow tract leading to the aortic valve. Abnormal anterior and apical displacement of the mitral valve encroached on the posterior and medial walls of the

FIG. 1.—Selective left ventricular cine-angiography of a 3-year-old boy with a small endocardial cushion defect, 30 per cent left-to-right shunt, trivial mitral incompetence, and mild additional coarctation of the aorta. (a) Antero-posterior view, early diastole: mitral valve is opening and shows the "goose-neck" deformity of the outflow tract (arrow). (b) Antero-posterior view, systole: pouting lips are formed by the superior and inferior halves of the cleft anterior cusp of the mitral valve (arrow). (c) Left anterior oblique position, systole: cleft mitral valve seen again; trivial mitral incompetence evident (arrow).

(a)

(d)

(b)

(e)

(c)

FIG. 2.—Selective left ventricular cine-angiography of a 5-year-old child with an endocardial cushion defect, cleft tricuspid and mitral valves, normal ventricular septum, and 70 per cent left-to-right shunt. (a) Antero-posterior view, diastole: the abnormal superior position of the cleft anterior leaflet of the mitral valve indents the outflow tract of the left ventricle, producing a typical "swan" or "goose-neck" deformity. The inferior part of the cleft cusp is also seen. The posterior leaflet is seen *en face* and is not distinct. (b) Antero-posterior view, systole: the abnormal contour of the left ventricle is shown. The normal ovoid left ventricle is divided into two parts: a rounded body with the mitral valve forming the right free margin, and an elongated narrow outflow tract. (c) Right anterior oblique, systole: the appearances are less specific and are not diagnostic. (d) Left anterior oblique, systole: abnormal pouting of the cleft valve and upper rim of the ventricular septum is again seen clearly. (e) Left anterior oblique, early diastole: "swan-neck" deformity seen again.

outflow tract, with narrowing and elongation of the lumen ("goose" or "swan-neck" appearance). The abnormal contour was demonstrated best during early diastole when a wave of non-opacified blood opened the mitral valve.

Another important finding was the mitral valve, which occupied an abnormal plane and position, with its deformed architecture clearly visible. In the normal subject, the valve is seen in the right anterior oblique position, but in every patient studied the antero-posterior position showed the valve profile best. We performed duplicate left ventricular cine-angiocardigrams in 5 patients to compare the antero-posterior and right anterior oblique positions. The antero-posterior position was always more useful. In health the mitral valve orifice forms only the upper two-thirds of the right free margin of the left ventricle, but in our patients most of the right free margin was formed by the mitral orifice. Thus, during diastole, a large front of non-opacified blood entered the ventricle defining the superior and inferior portions of the cleft anterior leaflet with the posterior leaflet seen partly *en face*.

The abnormal architecture of the cleft rolled valve was seen best during systole when the inferior and superior parts of the cleft anterior cusp bulged into the left atrium. In the milder deformities with minimal mitral incompetence, the junction of the two halves of the anterior cusp formed a small pocket (Fig. 1b). Increasing degrees of mitral incompetence were observed as a positive jet into the left atrium or directly into the right atrium through the endocardial cushion defect. Abnormal mobility of the valve with slight tethering of the cusps by the shortened chordae was also observed during diastole, while in systole it gave the valve a serrated appearance.

The atrial septal defect was shown only in the presence of mitral incompetence when the jet of contrast medium was directed through the left atrium into the right atrium.

A deficiency of the ventricular septum could not be assessed in the antero-posterior position.

Other radiographic positions were also used. The right anterior oblique position had no advantage over the much simpler antero-posterior position. The left anterior oblique position demonstrated the ventricular septal defect in 2 of 3 patients. In the third patient it was uncertain whether contrast passed across the mitral valve into the left atrium and thence into the right atrium and ventricle or whether direct passage occurred across the ventricular septal defect into the right ventricle.

DISCUSSION

Embryology. In the normal foetus three structures form the atrioventricular junction: the septum primum, the endocardial cushions, and the ventricular septum (Fig. 3). The septum primum grows downwards from the postero-superior aspect of the common atrial wall towards the atrioventricular canal, and divides the common atrial chamber into two halves, with the ostium primum as a residual interatrial communication at its lower margin. The endocardial cushions then grow into the lumen of the common atrioventricular canal and fuse, forming the right and left orifices (tricuspid and mitral). The cushions also arch in the shape of a U so that the opening points forwards and to the left and the septum primum fuses with the middle of the two cushions. The ventricles are bisected by the thick muscular interventricular septum, the upper margin of which fuses with the arched endocardial cushions. Thus, the left border of the arch faces the left *ventricle*, the right border faces the right *atrium*, and the tricuspid lies at a lower level than the mitral valve. The endocardial cushions form the septal leaflet of the tricuspid valve, the atrioventricular septum, and the anterior leaflet of the mitral valve. The U-shaped bay of the endocardial cushion is important as it helps to form the conduit through the primary inter-ventricular foramen, leading from the body of the left ventricle to the aorta.

In an endocardial cushion defect the normal U-shaped bay does not develop so that the septum primum is unable to fuse with the upper margin of the ventricular septum (Van Mierop *et al.*, 1962). The interatrial defect consists mainly of the absent atrioventricular septum but it becomes interatrial because of the apical displacement of the attachment of the cusps of the mitral valve. If the defect is large, part of the lower margin of the normal interatrial septum may also be absent. The lower margin of the defect always has a concave scooped-out appearance where the endocardial cushions are deficient and the normal atrioventricular septum fails to appear (Frater, 1965). The medial third of the mitral valve is attached to the crest of the septum and not to the normal atrioventricular septum, so that it is displaced forwards and towards the apex of the heart. This is responsible for the abnormal position and plane of the valve. The valve itself is frequently cleft (partially or completely) with shortened abnormal chordae attached to the crest of the septum instead of the antero-medial papillary muscle. The cleft divides the valve into a normal superior half and an abnormally situated inferior half. The atrioventricular septum normally forms the postero-medial wall of the outflow tract of the

FIG. 3.—Embryology of the endocardial cushions. (a) Early stage development. Septum primum (s.p.) has formed and divides the left and right atria. Development of endocardial cushions (e.c.) has started. (b) Endocardial cushions have started to arch to form the mitral (m) and tricuspid (t) orifices. Septum secundum (s.s.) and muscular ventricular septum (m) have also developed. (c) Fully mature foetus. Endocardial cushions have arched to form the atrioventricular septum, have joined the septum primum and membranous ventricular septum (ms) and the mitral and tricuspid valves. The mitral and tricuspid valves lie at different levels. The outflow tract of the left ventricle (o.t.) is bounded partly by the atrioventricular septum (dotted) and the aortic leaflet of the mitral valve. (d) Patient with an endocardial cushion defect: atrioventricular septum has not formed, so that the aortic leaflet of the mitral valve is abnormal and displaced downwards and is attached to the upper border of the ventricular septum. This encroaches on the outflow tract of the left ventricle which is narrow and deformed. The defect is shown (e.c.d.).

left ventricle, so that its absence, together with the abnormal origin of the mitral valve, disturbs the normal architecture of the outflow tract. The defects encroach on the postero-medial walls forming a characteristic deformed, narrow, and elongated outflow tract.

In the more severe abnormalities, the tricuspid valve may also be cleft and part or all of the ventricular septum may be absent.

Clinical Importance of the Abnormal Cine-angiogram. We have seen the abnormal configuration of the left ventricle only in patients with an endocardial cushion defect where the

characteristic pathology produces this unique radiological profile. This picture does not occur in a simple ventricular septal defect, a combined ventricular and secundum atrial septal defect, or a secundum atrial septal defect with associated mitral incompetence. It is also useful to identify the occasional patient in whom a secundum atrial septal defect is associated with left axis deviation of the electrocardiogram.

The route to the left ventricle is also of some importance. The tip of the angiographic catheter may be passed into the left ventricle via the septal defect. Care must then be taken to avoid catheter recoil during the injection of contrast

medium. The best and most consistent results are obtained when the catheter is advanced retrogradely from the aorta.

When left ventriculography is used as a routine, the left ventriculogram should be recorded in two planes: in the antero-posterior plane to demonstrate the abnormal outflow tract, position, and plane of the mitral valve and mitral incompetence; and in the left anterior oblique position to demonstrate the presence of an additional ventricular septal defect.

SUMMARY

Selective left ventricular cine-angiography was performed in 13 patients with an endocardial cushion defect. The diagnosis was confirmed at operation in 10. All 13 had an ostium primum atrial septal defect and cleft mitral valve, 5 had a cleft tricuspid valve, 3 had a ventricular septal defect, and 2 had additional pulmonary stenosis.

The findings on auscultation and the electrocardiogram were useful in establishing the correct diagnosis. Cardiac catheterization and multiple indicator dilution curves provided valuable additional confirmatory information, but the left ventricular cine-angiogram was the most helpful special investigation.

In the simple deformities (atrial septal defect with cleft mitral valve), the "swan-neck" deformity of the left ventricle, with an abnormal position, profile, architecture, and incompetence of the mitral valve, was demonstrated best in the antero-posterior position. The right anterior oblique position added little extra information but the left anterior oblique position was useful to demonstrate deficiency of the ventricular septum.

The radiological appearance was explained by the abnormal embryology and pathological anatomy.

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