The "Goose Neck" of the Endocardial Cushion Defect: Anatomic Basis*

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The "goose neck" deformity as seen in left ventriculograms in the endocardial cushion defect persists after surgical treatment in which the subaortic deficiency of the ventricular septum is built up with prosthetic material and the natural mitral valve is replaced by a prosthesis. Anatomic studies indicate that the diaphragmatic (inflow) aspect of the left ventricle and the ventricular septum are short relative to the outflow length of the ventricular septum. In addition, there is a deficiency in the subaortic aspect of the ventricular septum. The study suggests that the term "goose neck" deformity following correction of the deficiency of the ventricular septum appears to be related to the short nature of the inflow part of the left ventricular wall at its diaphragmatic aspect. The study suggests that the term "goose neck" deformity is better replaced by "goose" deformity, since the deficient posterior wall of the left ventricle outlines the curvature of the back of the "goose" which persists in spite of correction at operation.

Persistent common atrioventricular canal, the so-called endocardial cushion defect, is characterized by a defect in the lowermost portion of the atrial septum, clefts in the mitral and/or in the tricuspid valves and deficiency in the ventricular septum. Complete and partial varieties have been defined by Rogers and Edwards. In the complete form, clefts in the mitral and tricuspid valves are continuous and the atrioventricular valvular mechanism is represented by a common valve. The developmental aspects of the malformation and the functional characteristics of the cleft mitral valve were described by Van Mierop and Alley.

The partial form, sometimes called ostium primum atrial septal defect, is associated with a cleft in the mitral valve. Although deficiency in tissue of the ventricular septum and of the posterior wall of the left ventricle is present in each type, an actual interventricular communication usually is present only in the complete variety.

From the angiocardiographic point of view, the diagnostic pattern, as seen in left ventriculograms, is commonly called the "goose neck" deformity.

This appearance has been described as resulting mainly from the abnormality of the mitral valve. Gotsman et al. state that, "abnormal anterior and apical displacement of the mitral valve encroached on the posterior and medial walls of the outflow tract with narrowing and elongation of the lumen ('goose' or 'swan neck' appearance)." Others have also implicated abnormal positional attachment of the anterior mitral leaflet as the basis for the characteristic deformity as seen in left ventriculograms.

Our experience has been that the "goose neck" deformity persists after removal and prosthetic replacement of the mitral valve along with insertion of a patch to enlarge the ventricular septum. This experience, based on two patients on whom operation was performed, indicates that the basis for the...
"goose neck" deformity is more complex than abnormal position and attachment of the mitral valve. Our evidence favors the nature of the left ventricle as a whole as the factor yielding the so-called "goose neck" deformity of this malformation.

**Observations**

**Results of Operation in Two Cases**

In two patients (a four-year-old boy and a 12-year-old girl), each with the complete variety of the endocardial cushion defect, left ventriculograms before operation showed a characteristic "goose neck" deformity of the left ventricular outflow tract (Fig 1a).

Surgical repair was performed by a method previously described. By this method not only is the atrial septal defect closed, but in addition, a Teflon patch is secured to the superior edge of the deficient ventricular septum. Placement of the latter patch accomplishes several desired features as follows: (1) it allows closure of the interventricular communication without crowding the left ventricular outflow tract; and (2) it allows the base of the prosthesis replacing the removed natural mitral valve to lie at a higher level anteriorly than the abnormal level of the natural mitral valve. The latter step is carried out so as to avoid the production of left ventricular outflow obstruction by the prosthesis. Posteriorly, however, the prosthesis is secured at the level of the natural mitral annulus.

Each patient survived the operation, and left ventriculography was performed about one year later.

These studies showed persistence of the "goose neck" deformity of the left ventricular outflow tract (Fig 1b) but with somewhat less narrowing of the left ventricular outflow tract than seen before operation.

These observations conform to those of Griffiths et al, who described angiograms before and after operation in cases of suture repair of ostium primum defect with cleft mitral valve. They indicated that angiograms after operation frequently showed a change toward normal in the contour of the outflow tract, although clearly a "goose neck" deformity still existed in at least two of the cases they illustrated.

Since in our cases the outflow tract of the left ventricle had been widened by the placement of artificial septal material and the mitral valve had been removed, persistence of the "goose neck" deformity after operation suggests a more complicated basis for the deformity than previously explained.

The basis for the deformity appears to be related in part to the short nature of both the left ventricular diaphragmatic wall and posterior aspect of the ventricular septum which characterize this malformation. The features are elaborated on in the following.

**Measurements in Specimens**

Several measurements were made in a manner similar to those described by Goor et al of the lengths of the ventricular septum both in controls and specimens with the endocardial cushion defect. These measurements are named and defined below.

1. Inflow length: This is the length of the posterior (diaphragmatic) aspect of the left ventricular
base as measured from the apex to the mitral ring at the center of the posterior leaflet.

2. Outflow length: Two measurements were made in this area, one from the left ventricular apex to the upper aspect of the commissure between the left and right aortic cusps (L-R commissure) and the other between the apex and the upper aspect of the commissure between the left and posterior aortic cusps (L-P commissure). Since in any one heart the distance between the apex and one commissure was essentially the same as that to the other, one value is taken and designated as the outflow length.

Eighteen specimens showing persistent common A-V canal were studied. One was from an adult and the remainder were from children or infants. Measurements were made of ten control specimens of hearts considered to be normal. Seven were from adults, one from an infant aged six months, one from a newborn (one day) and one from a stillborn. The inflow length among the cases with the malformation varied in length from 22 to 56 mm in the children and infants, while in the one adult, inflow length was 85 mm. In the one adult with the endocardial cushion defect, the outflow length was 115 mm, while among the children and infants with this malformation, the length varied from 26 to 80 mm. In each case, a ratio was calculated of the inflow length to the outflow length. Assuming the outflow length to be 100 percent, the inflow length was calculated as a percentage of the latter. In specimens with the malformation, this varied from 70 to 93 percent in the children and infants, while in the adult it was 74 percent. The average figure for the length of the inflow tract was 77 percent of the outflow length. Among the controls, the percentage of the inflow length to the outflow length was as follows: in the stillborn, 95 percent; in the newborn, 100 percent; in the infant, 93 percent; and in each of the seven adults it was 100 percent, yielding a control average inflow length of 98 percent of the outflow length.

It is to be recognized from the foregoing that part of the deformity of the ventricular septum in the endocardial cushion defect, therefore, is one in which there is deficiency in the length of the posterior wall of the left ventricle, including the related ventricular septum (Fig 2).

**Comment**

In the endocardial cushion defect, in addition to deficiency in length of the diaphragmatic aspect of the left ventricular wall and of the posterior aspect of the ventricular septum, there is deficiency of the ventricular septum in the subaortic area. If, in the normal heart, one traces the upper edge of the ventricular septum from the aortic valve to the dia-
phragmatic wall, a straight line is followed. In the endocardial cushion defect, on the other hand, a concavity is obtained, ending at the deficient upper end of the diaphragmatic aspect (Fig 2).

In the outflow region of the left ventricle, the anterior aspect of the anterior mitral leaflet, in the partial form, or the anterior leaflet of the common A-V valve, in the complete, vary in their attachment to the superior edge of the deficient ventricular septum. In the partial type, usually the ventricular surface of the anterior half of the anterior mitral leaflet is fused with the crest of the deficient ventricular septum. The fixation of the valvular tissue precludes any significant change in the caliber of the outflow tract during the various phases of the cycle.

In the complete variety, the anterior leaflet of the common A-V valve is either not attached to the ventricular septum or attached by way of the chordae. In either case, the caliber of the outflow tract changes as the related A-V valvular tissue moves. During ventricular systole, upward movement of the A-V valvular tissue would widen the outflow of the left ventricular outflow tract. During ventricular diastole, the mobile A-V valve lies against the superior edge of the deficient ventricular septum (Fig 3).

It has been emphasized that the “goose neck” deformity is best brought out during ventricular diastole. This is understandable on the basis of the fact that in diastole the mitral mechanism (whether intimately attached or not attached to the ventricular septum) lies against the superior aspect of the ventricular septum and serves to outline the extent and shape of the septum and of the left ventricular cavity (Fig 2).

With this view in mind, it is understandable that there would be a narrow subaortic shadow during left ventricular diastole but left unexplained is the persistence of the “goose neck” deformity after the surgical procedures described. Included in the procedure is the addition of material to the subaortic part of the deficient ventricular septum. Also the natural mitral valve is removed and in its place the prosthetic valve anteriorly is secured more superiorly than the natural mitral valvular annulus. Even though the width of the left ventricular outflow tract and the “neck of the goose” is widened, the “goose neck” deformity persists. The part of the heart that is unchanged surgically lies in the diaphragmatic aspect. Here, the prosthetic valve is secured at the same level as the annulus of the mitral ring, and the operation causes no change in the short length of the diaphragmatic aspect of the left ventricle (Fig 4). These features must, therefore, play a role in the persistence of the basic angiographic deformity even though the “neck of the goose” has been widened. If one looks at the contour of the entire left ventricular shadow, posteriorly there is a concavity which might be likened to the “back of the goose.” It appears, therefore, that in this malformation the angiographic shape of the left ventricle is that of a “goose” rather than a “goose neck.” Thus, while the neck may be widened surgically, the shape of the diaphragmatic aspect of the left ventricle persists to yield the outline of the entire “goose,” the “neck” being part of the whole.

**Figure 3.** Diagrammatic portrayal of left ventriculogram in complete variety of endocardial cushion defect. (a) Diastolic phase. As anterior leaflet (A.M.) of common atrioventricular valve lies against superior edge of deficient ventricular septum, “goose neck” deformity is better brought out during diastolic phase than during systolic phase. (b) Systolic phase. Since anterior leaflet of common atrioventricular valve is attached by chordae to deficient ventricular septum, leaflet (A.M.) may move upward during systole. Posterior mitral leaflet (P.M.) also moves upward. Enlargement of outflow tract of left ventricle incident to upward displacement of atrioventricular valvular tissue enlarges left ventricular outflow tract during this phase and obscures “goose neck” deformity.
"GOOSE NECK" OF ENDOCARDIAL CUSHION DEFECT

It is apparent that among patients with the endocardial cushion defect not having operation there are variations from the classic "goose neck" deformity toward the normal. This conforms to our observation that the length of the diaphragmatic aspect of the left ventricle, while deficient in all, may, in some, approach normal. While in cases with the greatest degree of inflow shortening the length of the inflow tract was 70 percent of outflow tract, at the other extreme was a case in which the inflow tract was 93 percent as long as the outflow tract.

REFERENCES