Inverted Malformations in Corrected Transposition of the Great Vessels

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The anomaly known as corrected transposition has been adequately described.\(^1\)\(^2\) In essence, while the atria are normal, the great vessels are transposed and the ventricles and atrioventricular valves are inverted; that is, each of these structures assumes a mirror-image pattern of the opposite structure as seen in the normally oriented heart.

Within the normally oriented heart there are certain malformations that apply to certain specific structures. When corrected transposition is present, such malformations follow the structures from which they arise and therefore appear on the contralateral side of the heart from which they occur in the normally oriented heart. Such malformations occurring in hearts with corrected transposition may be defined as inverted malformations.

Appearing on the contralateral side of the heart from the usual, inverted malformations yield functional derangements that are different from those of comparable malformations occurring in the normally oriented heart.

Malformations that have been described in corrected transposition and that may be considered examples of inverted malformations include the Ebstein-like malformation of the left-sided atrioventricular valve,\(^3\)\(^4\) membranous subpulmonary stenosis,\(^5\) and accessory tissue of the subpulmonary area,\(^6\) the last condition being the inverted form of subaortic stenosis caused by accessory tissue of the mitral valve in the normally oriented heart.\(^7\)

The purpose of this report is to illustrate two additional inverted malformations as observed in corrected transposition. The first of these took the form of an anomalous muscle bundle of the arterial ventricle (fig. 1), a condition which is an inverted form of the condition commonly known as anomalous muscle bundle of the right ventricle.\(^8\) In this case the corrected transposition was of a special form in which, while the great vessels were transposed and the ventricles inverted,

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Figure 1
Case 1. A 3½-month-old baby girl. The arterial (left-sided) ventricle and the ascending aorta. At the junction of the inflow and outflow portions of the ventricle is a pyramidal-shaped mass of muscle (M.), which divides that part of the ventricle between the atrioventricular and aortic valves into two channels (one opened and one containing probe). The malformation observed in this example of corrected transposition is considered to be an inverted form of the malformation known as anomalous muscle bundle of the right ventricle in normally oriented hearts. In the instance shown the anomalous mass of muscle was responsible for subaortic stenosis.
the atrioventricular valves were not inverted.

The second of the two conditions to be shown represents inversion, in partial form, of the developmental complex described by Shone and associates. Those authors described, in normally oriented hearts, coexistence of four obstructive malformations as follows: (1) supravalvular ring of the left atrium,
(2) parachute mitral valve, (3) subaortic stenosis, and (4) coarctation of the aorta. In the case of corrected transposition representing the inverted form of this condition, the right-sided atrioventricular valve exhibited a parachute deformity and in the venous ventricle there was subpulmonary stenosis (fig. 2).

Summary and Conclusions

Inverted malformations are defined as those malformations that are specific for certain anatomic structures in the heart and that occur in corrected transposition. An inverted malformation appears on the contralateral side of the heart from that characteristic for the basic malformation when present in the normally oriented heart.

Two cases of corrected transposition with inverted malformations are presented. One was an example of an anomalous muscle mass occurring in the arterial ventricle of a heart with a special form of corrected transposition. This mass is considered to be an inverted form of anomalous muscle bundle of the right ventricle. The second case of corrected transposition was associated with a parachute deformity of the right atrioventricular valve, membranous subpulmonary stenosis, and bicuspid pulmonary valve. These malformations are considered to represent the inverted form of the developmental complex in which a parachute deformity of the mitral valve and subaortic stenosis are part.

References

Quiet Reverence

The way in which the persecution of Galileo has been remembered is a tribute to the quiet commencement of the most intimate change in outlook which the human race had yet encountered. Since a babe was born in a manger, it may be doubted whether so great a thing has happened with so little stir.—A. N. Whitehead.

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