Surgical Anatomy of the Coronary Artery Distribution in Congenital Heart Disease

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During the initial phases in the development of open-heart surgery, problems related to the perfusion procedure were common causes of morbidity and mortality. With improved perfusion techniques and increased knowledge of the physiology of cardiopulmonary bypass, such problems have become less frequent. Now that the perfusion procedure itself is accomplished with considerable safety, the surgical anatomy of congenital cardiac malformations has assumed increasing importance. The gross anatomic features of various cardiac deformities were elucidated largely before the advent of open-heart surgery, and the pertinent anatomy of the cardiac conduction system has been the subject of several recent studies. The coronary artery distribution in congenital heart disease has received scant attention, although recent reports attest to the importance of anomalies of coronary circulation encountered during open-heart surgery.

The present report deals with distribution of the coronary arteries in congenital heart disease, with emphasis on the two areas most frequently attacked during open-heart surgery: the right ventricular outflow tract and the region surrounding septal defects.

Materials and Methods

Studies were performed on 154 hearts with congenital malformations and 24 normal human hearts. Coronary arteries of fresh specimens were injected with barium-latex or lead-vinylite suspension. Roentgenograms were made of the unopened and opened hearts, and the specimens were subsequently cleared and photographed. Gross dissection of the major coronary vessels was performed on formalin-fixed hearts.

Results

The ventricular septum in normal and malformed hearts received a generous blood supply from branches of the anterior and posterior descending arteries (fig. 1). The artery to the atrioventricular node arose from the right coronary artery in approximately 90 per cent of cases and from the left coronary in the remaining 10 per cent. This artery originated at a sharp U-shaped turn of the right or left coronary artery near the coronary sinus.

In ostium primum defects the artery to the atrioventricular node lay in a vulnerable position beneath the inferior (dorso-caudal) margin of the defect (fig. 2). Secundum defects, however, were well removed from the blood supply to the conduction system (fig. 3). Most ventricular septal defects, including the ventricular component of atrioventricular canals, lay anterior (ventral) to the main artery to the atrioventricular node, although small vessels in the area of the bundle branches coursed in the ventricular septum immediately beneath the defects (fig. 4).

The right ventricular outflow tract in normal hearts was supplied by small branches of the anterior descending artery and aortic branches of the right coronary artery. The usual site of right ventriculotomy lay in an avascular area of the normal heart; however, in tetralogy of Fallot, marked increase in vascularity was observed in the right ventricular outflow tract. Less striking increases in vascularity were observed in cases of ventricular septal defect and isolated pulmonic stenosis.

Occasionally major coronary vessels were
Figure 1
Photograph and drawing of an injected and cleared specimen of the ventricular septum of a normal heart. The septum receives a generous supply from the anterior descending artery of the left coronary and the posterior descending artery from the right coronary. The artery to the atroventricular node arises at a sharp bend in the right coronary artery.

Figure 2
Photograph and drawing of an injected and cleared specimen with an ostium primum type of atrial defect. The artery to the atroventricular node is seen to course immediately beneath the inferior (dorso-caudal) rim of the ostium primum defect.

found to pass across the right ventricular outflow tract, in the area usually incised during right ventriculotomy. The anterior descending artery occasionally arose from the right rather than the left coronary artery (fig. 5). This anomaly was observed especially in cases of tetralogy with marked dextroposition of the aorta.
A variant of single coronary artery of particular importance was the single right coronary artery in which the entire left coronary branch passed in front of the pulmonary artery (fig. 6). A similar situation was observed in one instance in which the left coronary, after arising from the left sinus of Valsalva, passed in front of rather than behind the pulmonary artery (fig. 7).

In most instances of anomalous distribution of vessels in the area of the right ventricular outflow tract, external examination of the heart revealed the presence of a major vessel in an abnormal location. In occasional instances, however, a major coronary vessel was buried within the myocardium and would not have been apparent on gross examination at the operating table.

Discussion

Anomalies of the coronary arteries are generally considered of no clinical significance, except in such conditions as coronary artery origin from the pulmonary artery and coro-
Figure 5

Drawing of a heart showing the anterior descending artery arising from the right coronary artery. The anterior descending artery in this instance crosses the outflow tract of the right ventricle.

Coronary artery distribution

Coronary arteriovenous, arterio-atrial, or arterio-ventricular fistulas. In open-heart surgery, however, the distribution of major coronary arteries may assume crucial importance. The artery to the atrioventricular node is a constant vessel arising from the left or right coronary artery in the area of the coronary sinus. This vessel lies in a vulnerable position immediately beneath the inferior (dorsocaudal) rim of ostium primum defects. Smaller vessels, from both the right and left coronary arteries, accompany the bundle branches. Whether postoperative conduction disturbances are related to direct injury to the conduction system, to compromise of its blood supply, or to other factors remains conjectural.

There is no doubt, however, about the importance of anomalous distribution of the coronary vessels in the region of the right ventricular outflow tract. In the most dangerous forms, either the anterior descending artery or the entire left coronary artery passes across the right ventricular outflow tract.

Senning reported five cases with a major anomalous vessel in this region. In two cases the anterior descending artery originated from the right coronary artery and coursed across the infundibulum. He reported one case in which the entire left coronary arose from the right coronary and coursed across an area of infundibular stenosis within the myocardium. The patient died immediately following division of this vessel.

In a summary of cases of tetralogy treated by open repair, Kirklin and associates described four instances of anomalous vessels in the region of the right ventricular outflow tract. In one instance a large vessel, presumably the left coronary, arose from a single right coronary artery. Its division during ventriculotomy resulted in death. In three cases the anterior descending artery arose from the right coronary. In one of these cases
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Drawing of a specimen with tetralogy of Fallot in which the left coronary artery arose from a normal site in the aorta but passed anterior to the pulmonary artery.

Figure 7

The pertinent embryology of this malformation, elucidated by Spitzer and Lev, has recently been reviewed.

Summary

Distribution of major coronary arteries was studied in 154 congenitally malformed hearts and 24 normal hearts.

The artery to the atroventricular node constantly arose in the area of the coronary sinus from either the right or left coronary artery. In specimens with ostium primum malformations, this artery coursed immediately beneath the inferior (dorso-caudad) rim of the defect.

Anomalous distribution of the coronary arteries in the region of the right ventricular outflow tract poses a serious problem in open-heart surgery. The two most frequent variants were anomalous anterior descending artery arising from the right coronary, and single right coronary artery with the left coronary branch passing across the right ventricular outflow tract.

Such anomalies occur most frequently in transposition complexes and tetralogy with extreme dextroposition of the aorta. Fatalities have been reported following division of these vessels.

Although some anomalies of the coronary vessels may be recognized on inspection of the heart at the operating table, a major vessel may lie buried within the myocardium.

References

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tricular septal defect and right-left shunt (Fallot's tetralogy), Acta chir. scandinav. 117: 73, 1959.


Thomas Sydenham
1624–1689

He that designs to attain to the right understanding of any Art or Profession, usually chooses some Eminent Man of the Art to be his Guide and Pattern, by whose Directions and Example, join'd with a tolerable Capacity, and sufficient Diligence, he is, and is deem'd, at a stated Period, legally qualified for the exercise of the Art he professes. And this I Take to be the best and readiest way of attaining to the knowledge of any Art.

But seeing by the Custom of our Country, Physicians are educated another way, and chiefly improve themselves by Study and Books, it behooves them to make choice of such Authors as have been much conversant in Practice, and are generally accounted Candid and Sincere. In the first Rank of these I place this Author who was peculiarly disposed for Practice, and a Vein of Sincerity runs through his whole Works, a thing very rare in Physical Authors, especially in the Modern; and for this reason it was, he was so much calumniated by ill Men.

This Worthy Man was always busied in improving the Practice of Physick, even then when he was stepping on the Threshold of the other World, and like the Great Archimedes, would not suffer himself to be interrupted by any thing but resistless Fate.

He died in the Sixty sixth year of his Age; A great Age it was for one that had been closely besieged, near half the time with Opprobrium Medicorum, the Gout; which finding its frequent Efforts repell'd by his great Skill, called in its Auxiliary, the Stone, and together, with much a-do, they storm'd the tottering Tenement of Clay, when by reason of Age it was Sarche Tenible any longer.

He was born in Dorsetshire: His Father was a Gentleman of good Reputation, and a plentiful Estate. He was educated in All-Souls College in Oxford, and was Fellow of the same, and afterwards a Member and Ornament of the College of Physicians in London.

He was Religious, Loyal, Learned, of a Solid Judgment, and Sterling Honesty: But I shall say no more of him, his Book will be the best and most lasting Monument of his Fame.
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