The Holmes Heart—A Variant of Cor Triloculare Biaatriatum

Report of Case in a Child

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There are thirteen cases of cor triloculare biaatriatum noted in Abbott's series of 1,000 cases; only six of these 13 had the additional feature of a rudimentary ventricle from which coursed the pulmonary artery, the aorta, or both. These were included by Harley in 1958 in his classification of 31 collected cases with a rudimentary ventricle. Sharp and Heath each reported an additional case the following year. These 33 cases excluded those with tricuspid or mitral atresia in which a single ventricle was present and those in which the accessory chamber was a blind pocket leading to an atrial outflow valve.

There are only three reports of the variant of cor triloculare biaatriatum called the Holmes heart. Holmes in 1824, described a 22-year-old man whose heart had a single ventricle with an outflow chamber from which a pulmonary artery coursed and which did not have transposition of the great vessels. Drey et al., in 1938, reported the second case—a 14-year-old girl who was cyanotic from infancy and who died of a brain abscess. Brown reported the third case in 1950—a 10-year-old girl believed to have tetralogy of Fallot who died after an anastomotic operation.

A fourth case is here reported: a 12-year-old boy whose catheterization data are presented and in whom a diagnosis of Holmes heart was made at open-heart surgery.

Case Report

T.J., a 12-year-old Negro boy, was born on January 24, 1950, weighing 6 pounds. Because of difficulty in the initiation of breathing at birth, oxygen was used for 24 hours. During a 2-week hospitalization, the infant was noted to tire easily and was tube-fed. Frequent respiratory infections and feeding problems continued throughout infancy, and several hospitalizations were required for bronchopneumonia.

His first admission was at 18 months of age for dyspnea and distended abdomen. Physical examination revealed a hyperactive heart with a loud systolic murmur along the left sternal border accompanied by a thrill. A chest x-ray showed both right and left ventricular enlargement, and an electrocardiogram was compatible with a diagnosis of left ventricular hypertrophy. A retrograde carotid angiogram revealed a normal aortic arch and tributaries. The hemoglobin was 13.3 Gm. A diagnosis of acyanotic congenital heart disease was made, type unspecified.

Chronic wheezing and respiratory infections continued throughout childhood. Digitalization became necessary at 2 years of age; cyanosis was first noted at 5 years, and mild clubbing at 6 years.

In 1958 he was readmitted for right heart catheterization. An increase in oxygen content at the right ventricular level and peripheral arterial saturation of 82 at rest and 54 with exercise were noted. The pulmonary artery pressure was 17/8 mm. Hg, the right ventricle pressure was 75/7 mm. Hg, increasing with exercise to 107/6 mm. Hg. Thus, an approximate 60-mm. Hg gradient in systolic pressure was recorded from right ventricle to pulmonary artery. The left ventricular pressure was 104/8. A phonocardiogram showed a closely split second sound and a loud systolic murmur beginning at the onset of the first sound, extending through systole, maximal in mid-systole, and tapered before the second sound. The second sound and the systolic murmur were both accentuated at the pulmonic area and at the left sternal border at the fourth intercostal space. The impression after catheterization was atypical tetralogy of Fallot.

By 1961 the child was mildly dyspneic with any exertion. An electrocardiogram (fig. 1) revealed left ventricular hypertrophy with incomplete right bundle-branch block, which was confirmed by vectocardiogram. A right heart
Electrocardiogram at age 11 showing left axis deviation and left ventricular hypertrophy.

catheterization again showed an increased oxygen content in right ventricular samples and a systolic pressure of 100 mm. Hg. An infundibular chamber was entered and had a pressure of 31 mm. Hg, with 21 mm. Hg in the pulmonary artery. The slightly anterior position at which the catheter entered the aorta suggested corrected transposition with pulmonary stenosis and ventricular septal defect.

At angiography the catheter tip was in the left ventricle for the first injection, from which the flow of dye coursed anteriorly even though the ventricle was posterior. After the catheter was repositioned, another injection was made and was followed by some delay in the disappearance of the dye. The impression was that the injection had been made into the pulmonary outflow tract just under the pulmonary valve and the dye seemed to pass anteriorly to the aorta.

At 12 years of age, an angiogram ruled out transposition but injection into the pulmonary artery did not reveal a single ventricle. Open-heart surgery was scheduled with the intention of closing the ventricular septal defect and correcting the outflow stenosis. When the pericardium was opened at surgery, the heart appeared grossly normal, without distortion or displacement of the major vessels and some hypertrophy of what appeared to be the right ventricle; the pulmonary artery was slightly anterior to the aorta and coursed to the left. The atrium and venous return appeared to be normal. Finger palpation through the atrial appendage revealed a normal tricuspid valve, no atrial septal defect, and what was initially thought to be a hypertrophied right ventricle with an infundibular ridge. The presumed right ventricle had a pressure of 80/10 mm. Hg, which was approximately equal to systemic pressure, and the infundibular pressure was 20/10.

A transverse incision was made just above the central portion of the presumed right ventricle. Immediately inside the ventriculotomy was a muscular ridge that partially occluded the right ventricular outflow tract and was excised for better exposure of the chamber. The outflow tract was noted to contain a normal-appearing pulmonary valve. Immediately inferior to the incision was a solid wall of myocardium that formed the apex of this small ventricular chamber. At the upper medial end of this was a large, high, ventricular septal defect through which the aortic valve was seen. There was no other opening into this chamber. A finger was inserted through the ventricular septal defect, introduced into the right atrium through what seemed to be a normal tricuspid valve, then redirected and passed through what appeared to be a normal mitral valve into the left atrium.

In order to verify these findings, a right atriotomy was made to permit the passage of an instrument—and later a finger—through a normal-appearing tricuspid valve into the apex of the larger ventricle. The small right ventricular chamber could be entered only through the septal defect. The ventriculotomy was closed and the patient had an uneventful recovery.

The piece of excised myocardium had a smooth, yellow-white endocardial surface. Microscopic examination revealed hypertrophy of individual muscle fibers, muscle bundles of varying size, and minimal subendocardial fibrosis.

Discussion

The Holmes heart has contributed to the careers of both Sir William Osler and Maude Abbott, and was the basis for the association of these two great figures in cardiology.

In 1899, Maude Abbott, as the curator of the pathological museum at McGill, asked Osler to help her to identify an unclassified specimen. He promptly recalled the specimen as one which he had demonstrated to his students while professor at McGill, and informed her that the case had been reported by "Old Dr. Andrew Holmes." Abbott republished this case in 1901.

This association, crystallized by the Holmes heart itself, later prompted Osler to ask Dr. Abbott to write a section on congenital heart disease in his System of Modern Medicine. The first edition showed Abbott's chart of 412
cases of congenital heart disease, a revision in 1915 raised this to 631, in 1927 to 850, and in 1932 to 1,000 cases.

It is of interest also that Dr. Holmes performed the autopsy on his case at the Montreal General Hospital in 1823, at the age of 26 years, in the presence of three other founders of the McGill Medical School. The presentation of this case to the Edinburgh Medical-Chirurgical Society was a "first" for this man who later became Dean of the McGill Medical School.

In cor triloculare biatriatum, a rudimentary chamber may be present, which may characteristically lie either anterior and to the right of the atrioventricular valves, or anterior and to the left. This position is thought to depend on the direction of rotation of the bulbo-ventricular position of the embryonic heart. If the conus rotates to its usual position, a right-sided accessory ventricle appears, but, when it rotates to the left, the accessory chamber is left-sided.

The accessory chamber functions as a ventricle in that it forms an outflow tract either for the pulmonary artery, the aorta, or both. When transposition of the great vessels occurs in addition to the bulbo-ventricular rotation, the accessory chamber leads to an aortic valve. When there is a right-sided rudimentary ventricle with normal position of the great vessels, it fits the description of the Holmes heart (fig. 2) and of our patient.

Rogers and Edwards reported a case of single ventricle with a pulmonary outflow pocket separated from the ventricle by a muscular ridge which offered little resistance to blood flow. Castrovinci and Cucci classified this case as a Holmes heart, although it was not reported as such.

Holmes did not consider the accessory ventricle as an outflow pocket nor the opening into it as a muscular ridge. He described the flow of blood from the left to the right ventricle through a 1/2 by 3/4 inch opening with tendinous margins just below the aorta. These measurements are comparable to the opening in our case (2.3 cm. diameter). Abbott in her re-examination of the Holmes heart stated that the opening was diamond-shaped and nonvalvular and just below and to the right of the aortic valve but above the tricuspid valve. The accessory chamber was described as triangular and at the right upper anterior angle of the heart. The muscular septum measured 1/2 inch in thickness. The diamond-shaped opening to it from the main ventricle was at its inner end. The normal-appearing pulmonary artery in front of that vessel was slightly larger than the aorta and coursed from right to left.

Our case differs from that of Rogers and Edwards in that the small right ventricle has a lower than systemic systolic pressure. The role of the crista supraventricularis in forming the aberrant septum was discussed by Rogers and Edwards. Others considered that the ridge forms from the muscle at the bulbo-ventricular junction.

The symptoms associated with Holmes heart, in general, include cyanosis, which may appear in infancy or, as in our case, not until midchildhood; clubbing; minimal exercise tolerance; and underdevelopment, as in Drey's case and ours. Holmes also found palpitation, dyspnea, and precordial pain, as well as anxiety, cough, and bloody mucus (these

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*Diagram showing relationship of great vessels and rudimentary chamber to the single ventricle and atrioventricular valves in the Holmes heart.*

*Figure 2*
symptoms may have been manifestations of a subacute bacterial endocarditis, which may have hastened death).

The murmur in the Holmes heart is systolic and harsh or blowing, probably reflecting the presence of pulmonary outflow pressure gradient or the sound of blood flowing through the septal defect into the rudimentary ventricle. The quality of the second heart sound was recorded previously only by Drey,6 who noted that it was "fairly loud." In our case, the second sound was closely split but not prominent.

The rudimentary ventricle in the Holmes heart probably acts as an infundibular chamber. The systemic pressure of the single ventricle is cushioned and hypertension may not develop in the pulmonary circulation. This rudimentary ventricle was the one probably entered at catheterization of our patient in 1961. The pressure was 31/5, while the pull-back pressure, probably of the single ventricle, was 100/5, and the pulmonary artery 21. In spite of this decrease in pressure, the pulmonary blood flow was not diminished (4.4 L./min.); the pulmonary-to-systemic flow ratio was calculated as 1.1/1. Holmes5 and Drey's6 cases died prior to the catheterization era, and the data on Brown's case7 are not published.

A clinical similarity exists between the Holmes heart and tetralogy of Fallot, but confusion exists for the following reasons: (1) in both, the pulmonary artery segment on the chest x-ray is likely to be small; (2) in the Holmes heart cyanosis is also not usually present at birth but develops in infancy or childhood; (3) the murmurs in the Holmes heart are compatible with a ventricular septal defect and pulmonary stenosis. In addition, the catheterization data show mixing of blood at the ventricular level and ready access to the pulmonary artery and the aorta from the ventricle. The presence of an infundibular chamber may also occur in tetralogy of Fallot; however, the important clinical distinction seems to be the electrocardiogram.

In Brown's case5 and in ours, a consistent left ventricular pattern was demonstrated. The additional findings of left ventricular hypertrophy and left axis deviation on the vectorcardiogram, not previously recorded in a Holmes heart, are also important, since at one time they were thought to be compatible with corrected transposition with ventricular septal defect.

What does heart surgery have to offer a patient with a Holmes heart? The creation of an artificial interventricular septum would be helpful if it could effectively divide the right from the left side of the heart. Since this is not possible with present technics, what other approach is available? If the opening between ventricles is too small, enlargement would increase pulmonary blood flow but there may be additional outflow tract obstruction, as in our case. Resection of this band of muscular tissue may or may not be beneficial. A Holmes heart associated with valvular pulmonary stenosis could conceivably benefit from a valvulotomy, but complete mixing of blood in the single ventricle might negate any relief offered by a Blalock or Potts anastomosis.

This variation of cor triloculare biventriculatum is extremely rare; four cases have been reported without being diagnosed prior to autopsy or surgery. Even though our case had three separate angiocardiograms and two complete catheterization studies, a diagnosis was not made prior to surgery and, as with other types of single ventricle, the prognosis is guarded.

Summary

A fourth case of Holmes heart is described in a 12-year-old boy, and developmental and corrective considerations are compared and discussed.

The first catheterization data in this condition are presented.

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The Functional Approach

It is with the symptoms of disease that the patient, and that the doctor mainly, contends; and the symptoms of heart disease may be said to derive almost exclusively from faults in function. Therefore, in managing our patients, our thoughts must be chiefly set in terms of function and not of structure. To whom I fail to teach this first simple, but essential, lesson I have naught to teach.—Sir Thomas Lewis. Diseases of the Heart. New York, The Macmillan Company, 1933, p. vii.
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