The Conotruncus

II. Report of a Case Showing Persistent Aortic Conus and Lack of Inversion of the Truncus

(A Bulboventricular Heart)

By DANIEL A. GOOR, M.D., AND JESSE E. EDWARDS, M.D.

SUMMARY
A primitive form of double-outlet right ventricle is described clinically and anatomically. A long tubular conus, arising exclusively from the right ventricle emptied into the transposed great vessels. Together with the presence of A-V canal malformation this heart represented a developmental arrest in the embryonic stage of the bulboventricular loop seen in horizon XV. Diagnosis is made on the basis of right ventricular angiography and theoretically, if this lesion is isolated, this conus malformation is surgically correctable.

Additional Indexing Words:
Right ventricle double outlet Transposition of great arteries Conus
Truncus Heart embryo

IN THE NORMAL development of the embryonic heart there are two critical processes which are involved in determining the definitive relationship between the aorta and the pulmonary artery. These processes are the shortening of the aortic conus and the inversion of the conotruncus. The purpose of this communication is to present the clinical and pathologic pictures of a previously undescribed pathologic condition, in which neither inversion (nor reverse torsion) of the truncus nor shortening of the conus takes place.

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Supported in part by Public Health Service Research Grant 5 R01 HL05694 and Research Training Grant 5 T01 HL05570 from the National Heart and Lung Institute.

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Received August 31, 1971; revision accepted for publication March 20, 1972.

Clinical History
A baby girl was 23 months old at the time of her death. Cyanosis was first noticed at the age of 6 weeks. Cardiac murmurs, cardiomegaly, and congestive heart failure were first detected at the age of 8 weeks. The patient suffered from recurrent episodes of pulmonary infections, and she was digitalized at the age of 3 months. Asplenia was diagnosed at the age of 3 months.

The physical examination revealed moderate cyanosis. There was a systolic thrill along the left sternal border as well as a right ventricular heave. Pulmonic closure was accentuated, and the second sound split widely and varied slightly with respiration. There was a grade IV/VI systolic regurgitant murmur along the left sternal border. There was an early systolic click and a grade II/VI diastolic murmur also along the left sternal border.

The electrocardiogram showed a first-degree heart block, right ventricular hypertrophy, and large P waves in V1, consistent with right atrial enlargement. Chest X-ray showed marked pulmonary hypervascularity, and cardiomegaly with the configuration of both transposition and anomalous pulmonary venous return.

Laboratory studies were not remarkable, with a hemoglobin of 15.4 g%.

The first catheterization and the angiographic studies were done when the patient was 3 months old (table 1). Angiography was done by injecting
Table 1

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<th>Catheterization Studies</th>
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the dye into the left ventricle (fig. 1). The dye passed through a VSD into a large conus, which showed marked dilatation and contraction during the cardiac cycle. (The term "conus" is used on the basis of the pathology found in the heart specimen. According to the original description given by Drs. E. Berry Hey and Richard Pyle from the University of Pennsylvania, the term "large unidentified structure" was used instead of conus. Otherwise the description of the angiography is based on their original report.) There was a delay in visualization of both great vessels, which visualized simultaneously. The pulmonary artery was strikingly far downstream with a long segment of heart, the conus, intervening between the great arteries and the ventricles. Because of the poor quality of the angiography and the bizzare picture that was presented, recatheterization was attempted at which the patient expired.

Autopsy showed the girl to be asplenic. The liver was in the midline and symmetric in appearance. The lungs were symmetric, each consisting of three lobes.

The heart was enlarged (figs. 2, 3). The apex pointed toward the left, and the aorta was in front of and to the right of the pulmonary artery.

The ventricular portion of the right chamber showed a well-developed right ventricle. The junction between the infundibulum and the ventricle was demarcated by the cephaloanterior border of the VSD. The length of the ventricle, as measured from the infundibuloventricular junction to the apex, was 3 cm. This was equal to the length of the infundibulum as measured from the infundibuloventricular junction to the semilunar valves. The infundibulum was the shape of a long muscular tube. The interior of this infundibulum was trabeculated. Distally (downstream), the infundibulum was continuous with the aorta and the pulmonary artery. At the distal portion of the infundibulum (or conus) there was a septum which represented the embryonic conus septum. This septum intervened between the subaortic and subpulmonary conuses which were equal in size. The length of the conus septum as measured along the blood flow was 0.5 cm, and it appeared grossly muscular. Distally, this septum was continuous with the septum aortopulmonale, which was intact. Anteriorly the conus septum was inserted into the anterior infundibular wall, and it was continuous with a distinct smooth ridge. Compared to embryos of horizon XV, this ridge corresponds to the distal end of conus ridge 3, and it measures about 1 cm in length. Posteriorly the distal conus septum was inserted into the posterior infundibular wall, and it was continuous with a heavy muscular ridge which ran initially along the posterior wall and then along the upper margins of the VSD to a point in front of and above the VSD. This ridge showed an identical distribution to that of the embryonic conus ridge no. 1. The portion of conus ridge 1, which was in front of and above the VSD, appeared in the left ventricular view as part of the anterior septum. The aortic valve was on the right and slightly to the front of the pulmonary valve. One coronary artery arose from the anterior aspect of the aorta. Before becoming a right arch, two brachiocephalic arteries originated from the aorta.

The pulmonary valve was slightly narrow, and it was bicuspid.

Other characteristics of this specimen included the following: the pulmonary veins drained via a common pulmonary vein into the right superior vena cava. (The pulmonary venous anomaly of this specimen was reported separately.3) There was a common atrium without anatomic characteristics of either right or left atrium. A single narrow band representing vestigial secondary atrial septum transversed in the anteroposterior direction through the common atrium.

There was persistent left superior vena cava, and it drained into the left half of the atrium. The right superior vena cava drained into the right half of the atrium. There was no coronary sinus.

There was a common A-V canal malformation with the common A-V valve being related to both ventricles.

Discussion

This specimen differed from the ordinary double-outlet right ventricle in two respects:
1. The conus was much longer than usual.
2. The distance between the conus septum (parietal band,4 muscular crest5) and the VSD was much longer than in the ordinary double-outlet right ventricle.

Surgically, so long as this conus anomaly is associated with A-V canal malformation, it is at present not correctable. If, however, a case...
of isolated conus malformation of this type is encountered, two possible surgical repairs should be considered: (1) diversion of the left ventricular stream into the aorta using a very long, $180^\circ$ spirally shaped patch; or (2) a long flat patch resulting in complete transposition of the great vessels with the addition of a Mustard procedure at the atrial level, a procedure used by others for Taussig-Bing malformation.\textsuperscript{6}
Figure 2

Photographs of the heart specimen. (A) Right ventricular view. (B) Left posterior atrial view. (C) Left ventricular view. The black marker in A protrudes from the pulmonary artery. The atrial view shows the common atrium with a vestigial atrial septum and the common A-V valve. IVJ = infundibuloventricular junction along the dashed line; RV = right ventricle; LV = left ventricle; Ao = Aorta.

Diagnostic Clinical Implications

It was stressed elsewhere that there is a high incidence of association between asplenia and bilateral conus. It is also accepted that asplenia is usually associated with other manifestations of symmetric development of viscera including the liver and the lungs. The presence of symmetric development of otherwise asymmetric viscera, in general, indicates that a bilateral conus, as in this case, might be present. While in the presence of a bilateral conus of the Taussig-Bing type (LV flow into the pulmonary artery) there is intense cyanosis from birth; in the present case, cyanosis was not noticed until she was 6 weeks old.

The demonstration of the long conus in angiocardiography is diagnostic. Despite the poor quality of the films this conus was
accurately described in the original catheterization report of this case. Two additional angiographic characteristics for this anomaly were the delay in visualization of the great vessels and the high position of the origin of the great vessels.

Developmental Significance
This heart showed an amazing similarity to the embryonic heart of horizon XV.1 In this horizon the aortic conus is almost as long as the right ventricle, and the pulmonary truncus is lying closer to the A-V canal than the aortic truncus. These two features are also seen in this specimen. The length of the aortic conus, and the relationship between the pulmonary and aortic valves, indicate that the normal truncal inversion, the conus absorption, and the leftward shift of the conoventricular flange that occurs during horizons XVI–XIX1, 2 do not occur in this specimen. Additionally, the A-V canal in this specimen remained as open as that in horizon XV.

Acknowledgment
The clinical information was kindly made available for this report by Dr. J. C. Shelburne, Director, Cardiac Catheterization Laboratory, Hospital of University of Pennsylvania. The authors would also like to acknowledge Miss Linda Field for her typing assistance in the preparation of this manuscript.

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Figure 3
Semidiagrammatic key illustration for figure 2.
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_Circulation_. 1972;46:385-389
doi: 10.1161/01.CIR.46.2.385

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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