CLINICOPATHOLOGIC CORRELATIONS

Bicuspid Aortic Valve
Comparison of Congenital and Acquired Types

By BRUCE F. WALLER, M.D., JOHN B. CARTER, M.D., HUGH J. WILLIAMS, JR.,
KUHYUN WANG, M.D., AND JESSE E. EDWARDS, M.D.

SUMMARY

Two dominant types of congenital bicuspid valves are described. The classical type is characterized by the presence of a low ridge or raphe along the aortic aspect of the conjoined cusp. The other is characterized by a tall raphe, the upper edge of which corresponds with the upper level of the aortic cusps. Some such ridges may result from acquired fusion of the adjacent halves of two cusps (yielding an acquired bicuspid valve). In other cases, the ridge is a protrusion of the aorta and not derived from fused cuspid tissue. Such valves are considered to portray a condition which may be termed pseudoacquired congenital bicuspid aortic valve. The acquired bicuspid valve in some cases is compounded of this congenital process and acquired fusion of cuspid tissue. The ratio of classical congenital bicuspid to pseudoacquired congenital bicuspid aortic valve is 4 to 1. Exceptional forms of pseudoacquired congenital bicuspid aortic valves are also described.

Additional Indexing Words:
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Commissures of aortic valve
Acquired bicuspid aortic valve

WHEN A BICUSPID aortic valve is encountered, the question as to whether it is congenital or acquired in nature is not always simple to determine. This is true in spite of the fact that the subject has been under study by a number of workers for many years. In the bicuspid aortic valve, one of the two cusps is larger than the other. The larger, the so-called conjoined cusp, exhibits a ridge or raphe along its aortic aspect. In some cases, the raphe does not extend upward to the level of the free edge of the cusp (figs. 1 and 2a-c). As the ridge is shallow, the conjoined sinus shows minimal subdivision into two parts. In this setting, a congenital basis for the bicuspid state is generally accepted and the process may be called classical congenital bicuspid aortic valve.

Problems in distinction arise when the upper edge of the ridge or raphe of the conjoined leaflet is composed of fibrous tissue and corresponds in height with that of the upper edges of the aortic cusps. Such a raphe may be termed a tall raphe or ridge (fig. 2d-f). In some cases, the tall raphe represents an acquired fibrous union of adjacent hemicusps.* In such cases, it can be demonstrated that the fibrous ridge is, in fact, formed by two adjacent hemicusps having become fused by fibrous tissue yielding the so-called acquired bicuspid aortic valve. There are, on the other hand, cases in which a tall ridge is not derived from fusion of two hemicusps. That such fibrous ridges are of congenital nature is supported by their composition,

*In closure of the aortic valve, one-half of a cusp makes contact with half of a second cusp while the other half makes contact with one-half of the third cusp. Thus, it is appropriate to divide each of the aortic cusps, including conjoined cusps, into two hemicusps.

From the Department of Pathology, United Hospitals-Miller Division, St. Paul, Minnesota and the Departments of Pathology and Medicine, University of Minnesota, Minneapolis, Minnesota.

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Address for reprints: Jesse E. Edwards, M.D., Department of Pathology, Miller Division, United Hospitals, 125 W. College Avenue, St. Paul, Minnesota 55102.

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may be termed *pseudoacquired congenital bicuspid aortic valves*.

These conclusions are reached after a study of aortic valvular commissures, both from adults and from infants, the latter with coarctation of the aorta. Some of the commissures studied were normal upon gross inspection, while others exhibited a tall ridge. For each case, a cross section was taken of the ascending aorta at a commissural area, the section to include the related cusp tissue as well as the ridge, if present. Cases with significant calcific disease were not included, since such changes obscure basic features which one needs to establish fundamentals.

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**Figure 1**

Two examples of classical congenital bicuspid aortic valve. In each, running from the aorta onto the aortic surface of the conjoined leaflet, is a low raphe (R). The upper edge of the raphe does not rise to the level of the free edge of the aortic cusps. (a) The raphe is particularly low and becomes indistinct as it reaches the conjoined cusp. (b) The raphe is taller than in (a) but does not rise as high as the free edge of the conjoined cusp.

which does not show elements of two cusps, and by the fact that this process may be seen in infancy. Such bicuspid valves are of congenital nature and...
Observations

The observations in the adults and in certain of the infants revealed recurring forms which may be termed standard patterns. In some instances, exceptional patterns were observed; these will be described in a separate section.

Standard Patterns

Infants with Coarctation

In infants with coarctation when the commissure was not unusual grossly, histologic examination showed that each of the two hemicusps forming the commissure were independent of each other to the point at which they joined the aorta. The union with the aorta was through a fibrous protrusion attached to the intimal aspect of the vessel. This protrusion is termed the commissural mound and corresponds with the mound seen at venous valves.1 At commissures which appeared normal, the mound projected only about one-sixth or less of the distance from the aortic wall to the center of the aortic lumen (fig. 3). In some cases, the mound was long and extended as far as the center of the aortic lumen (figs. 3c and 4).

Histologic examination of the mound showed it to be composed of fibrous tissue without signs of its being formed by fused hemicusps. The long mound corresponded to the tall ridge noted grossly. Grossly, the ridge might be uniform or it might show two parallel subdivisions, a picture which could suggest fusion of two adjacent hemicusps. Bicuspid valves associated with a tall fibrous ridge that could be interpreted as a long commissural mound were classified as pseudoacquired congenital bicuspid aortic valves. It is significant that the sinus related to the conjoined cusp was subdivided into two distinct components which in aortograms might be read as two distinct sinuses.

Adults

Aortic commissures from adults demonstrated the same features as described for infants with coarctation including variations in the length of the commissural mound (figs. 5–7). In addition, in some instances, fusion between adjacent hemicusps was

Figure 3

Aortic commissures in infants with coarctation of aorta. Photomicrographs of horizontal sections through the aortic wall and the commissural area as well as the two hemicusps contributing to the commissure. (a) A shallow commissural mound (M) receives the two hemicusps (C). (b) The commissural mound (M) is somewhat longer than in (a). (c) A long commissural mound (M) attaches to the aorta. At the luminal side of the mound are attached two related hemicusps (C). Elastic tissue stain; × 6.5.

Figure 5

Normal aortic commissures in adults. (a) Close-up view of gross specimen from which the photomicrograph is shown in (b). (b) Two hemicusps (C) are attached to a commissural mound (M). Elastic tissue stain; × 6. (c) Gross specimen. At the commissural area is a prominence (M) to which the two related hemicusps (C) attach. The prominence represents a relatively long commissural mound shown in (d). (d) Photomicrograph from gross specimen illustrated in (c). Commissural mound (M) is of moderate length. Two aortic hemicusps (C) attach to it. Elastic tissue stain; × 6.
Figure 4
Bicuspid aortic valve in an infant with coarctation of the aorta. (a) There is a long ridge representing a long commissural mound (M) to which, on the luminal side, are attached two hemicusps (C). Note that the ridge or commissural mound is cleft, suggesting two cusps. (b) Photomicrograph across the aorta and the commissural mound (M). The latter is long and uniform. The two related hemicusps (C) attach to the free end of the long mound. Elastic tissue stain; × 18.
added to the commissural mound and served to elongate the ridge (figs. 8–10). This means that in some instances of acquired bicuspid valve the fibrous ridge was composed in part of fused hemicusps and in part of an unusually long commissural mound.

**Exceptional Forms**

Most of the exceptional forms to be presented were purposefully taken from infants or children with coarctation of the aorta so as to support the congenital nature of the valvular abnormalities.

One of the exceptional forms was designated as
fenestration of cuspid tissue beneath tall fibrous ridge. This was characterized by a fibrous strand extending from the aorta to the upper aspect of the conjoined cusp, while a fenestration was present beneath the strand. In addition to observation of this condition in young subjects with coarctation (fig. 11a), we observed this process in the aortic valve of an 81-year-old man without other congenital anomalies (fig. 11b). A similar process was observed in the pulmonary valve of a patient with the tetralogy of Fallot (fig. 11c). The type of aortic valve under consideration represents a potential for aortic insufficiency as a consequence of rupture of the strand-like fibrous ridge. Such a condition in an adult was described from this laboratory.2

A second exceptional form might be called bridging of cuspid tissue across insertion of bifid ridge. This is characterized by two fibrous ridges or a bifid ridge extending from the aorta to the conjoined leaflet. A bridge of fibrous tissue extends between the cuspid insertions of the two ridges. This yields a picture suggesting a minor form of a third cusp (fig. 12).

Fenestrations of conjoined cusp associated with low insertion of fibrous ridge is characterized by two fenestrations being present in the opposing halves of the center of the conjoined cusp. A fibrous

Figure 7
Photomicrograph of aortic commissural area from an adult. A long commissural mound (M) lies between the aorta (A) on one side and two hemicusps (C) on the other. The mound appears similar to that seen in the infant illustrated in figure 4. Elastic tissue stain; × 6.
Photomicrographs of aortic commissural areas from adults. In each, the commissural mound is longer than in the commissures illustrated in figure 8. (a) Commisural area shows a long, fibrous ridge (M). (b) Fibrous ridge is composed of a moderately long commissural mound (M) in addition to fusion (between arrows) between adjacent hemicusps (C). Elastic tissue stain; × 6. (c) Unopened bicuspid aortic valve. A long, fibrous ridge (between arrows) extends onto the aortic surface of the conjoined cusp. (d) Photomicrograph of identified commissural area in (c). A long, fibrous mound (M) is responsible for the major part of the ridge. Some fusion (arrow) of the adjacent portions of the related hemicusps (C) is also present, as a contributing factor to the formation of the ridge. Elastic tissue stain; × 6. The valve illustrated in (c) and (d) is basically a pseudoacquired congenital bicuspid valve with minimal fusion between hemicusps.
An acquired bicuspid aortic valve in an adult. (a) Gross specimen of unopened aortic valve viewed from above. Fusion is major at commissure I, while some fusion is present at commissure II. Commissure III is not fused. (b) Photomicrograph of aortic valve shown in (a). Each commissural mound is short. There is fusion between the hemicusps at commissures I and II. Elastic tissue stain; × 3.5.
Figure 11
Fenestration of cuspid tissue beneath long fibrous ridge.
(a) From an 8-year-old boy with coarctation of the aorta. The aortic valve shows a bicuspid nature with a tall, fibrous ridge extending from the aortic wall onto the conjoined cusp. Probe is in a fenestration inferior to the ridge. The intact ridge tends to prevent prolapse of the conjoined leaflet.
(b) From an 81-year-old man without congenital anomalies. The aortic valve shows a fenestration (curved arrow) beneath a tall, fibrous ridge (between arrows). (c) Pulmonary valve from a 26-year-old man with tetralogy of Fallot. There is a tall ridge beneath which a defect (probe) is present. The picture is essentially similar to that shown in (a) and (b).
ridge extends from the aorta to the center of the conjoined leaflet at a level corresponding with the lower aspects of the fenestrations (fig. 13).

Isolated fibrous strand inserting into one cusp of a tricuspid aortic valve was observed in a patient with coarctation of the aorta and ventricular septal defect. Basically, this process is like that described above as fenestration of cuspid tissue beneath tall fibrous ridge. If one considers the involved cusp a conjoined cusp, the valve, in reality, is an example of a quadricuspid valve (fig. 14), while superficially resembling a tricuspid aortic valve.

Relative Incidence of Classical and Pseudoacquired Congenital Bicuspid Aortic Valves

In order to determine the relative incidence of the classical and pseudoacquired congenital bicuspid aortic valves, 72 hearts of infants under one year of age with aortic coarctation were studied. Of these, 40 (55 percent) showed a bicuspid valve. Of the 40 bicuspid valves, 32 (80 percent) were of the classical congenital type, while in eight cases (20 percent) the conjoined cusp of the bicuspid valve showed a tall ridge yielding a picture of pseudo-acquired congenital bicuspid aortic valve.

Comment

This study showed that the attachment of aortic cusps to the aortic wall is to a specialized fibrous mound located at the intimal aspect of the aorta. The mound varies in length. When it is long, it may extend as far as the center of the aortic lumen at which point two hemicusps attach. The latter...
WALLER ET AL.

Figure 13

Fenestrations of conjoined cusp associated with low insertion of fibrous ridge. Fenestrations are present in opposing halves of conjoined aortic cusp (C) while a fibrous ridge (R) inserts below the fenestrations. (a) From a 28-year-old man with coarctation of the aorta. (b) From an adult with congenital polycystic kidneys.

picture is one of a bicuspid valve in which the two hemicusps form a conjoined cusp.

In adults, one sees bicuspid aortic valves with tall ridges which have one of two compositions. One process is exactly like that in certain infants and may be considered to be congenital, while the other is represented totally or in part by acquired fusion of two cusps.

When two hemicusps attach to a long mound, greater tension at the sites of attachment occur than when the mound is short. Thus, there appears to be a greater tendency for fusion between hemicusps when the mound is long. Thus, the tall ridge of some acquired bicuspid aortic valves is compounded of a long commissural mound plus fusion between the two related hemicusps.

Regarding the relative incidence of the classical and pseudoacquired congenital bicuspid valve, our studies indicate a ratio of 4 to 1.

It is to be emphasized that in both acquired and pseudoacquired congenital bicuspid valves the sinus related to the conjoined cusp is divided into two distinct subdivisions. Thus, in aortography, demonstration of three sinus formations does not exclude the presence of a bicuspid aortic valve.

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