Clinical Anatomy of the Normal Pulmonary Root Compared With That in Isolated Pulmonary Valvular Stenosis

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Objectives. This study aimed to clarify the clinical anatomy of the pulmonary root.

Background. Many descriptions of valvular anatomy have focused on the annulus, leading to varied interpretations of abnormal valves.

Methods. Twenty-two heart specimens with isolated pulmonary valvular stenosis were examined to analyze the gross structure of the pulmonary root. For comparison, we examined a normal series of a similar age range together with nine adult hearts. Serial histologic sections were prepared from five specimens.

Results. The normal pulmonary valve is enclosed in a proximal sleeve of free-standing right ventricular infundibulum supporting the fibroelastic walls of the pulmonary sinuses at the anatomic ventriculoarterial junction. The valvular leaflets are attached in semilunar fashion across this junction, delimiting the extent of the valvular sinuses. The stenotic valves were separated into dome-shaped valves, dysplastic valves and a third group of less typical cases. In the dome-shaped valves, which had a relatively circular origin of their leaflets, three raphes were tethered to the arterial wall at the sinutubular junction, producing a waistlike narrowing. The leaflets of the dysplastic valves were attached in a relatively normal semilunar fashion, but stenosis was caused by thickening of the leaflets at their free edges. Serial histologic sections through normal and abnormal valves failed to demonstrate any well-defined fibrous “annulus” that could be of clinical relevance.

Conclusions. Unlike the normal and the dysplastic valves, the dome-shaped valves have circular rather than semilunar lines of attachment of the valvular leaflets. Liberation of the fused zones of apposition of the leaflets within the dome is unlikely to restore such abnormal valves to normal structure, even if this procedure relieves the stenosis.

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another, the pulmonary root had been excised and replaced by a homograft. The latter two cases, together with one mutilated specimen, were excluded from further examination. For comparison with normality we examined 22 hearts from children of comparable age (1 day to 3 years, median 124 days), together with nine normal adult hearts.

All hearts were analyzed macroscopically, paying special attention to the structures of the pulmonary root. In five selected specimens, we prepared serial sagittal and transverse sections of the pulmonary root. The histologic sections were stained with elastic van Gieson stain, which provided differentiation between myocardium, elastic and fibrous tissues.

Definition of terms. Using the principles for our definition of structures of the aortic root (4,5), we describe the pulmonary root as that part of the right ventricular outflow tract that supports the leaflets of the pulmonary valve. It is limited distally by the sinutubular junction. Its components are the sinuses of Valsalva, the valvular leaflets, the interleaflet triangles and the free-standing distal right ventricular muscular infundibulum (Fig. 1).

The sinutubular junction separates the pulmonary valvular sinuses from the tubular component of the pulmonary trunk. It also is the level of insertion of the peripheral ends of the zones of apposition between the leaflets (the so-called commissures) into the arterial wall.

The sinuses of Valsalva are the three parts of the pulmonary root confined proximally by the semilunar attachments of the valvular leaflets and distally by the sinutubular junction. According to their relation to the aorta, we distinguished among left-facing, right-facing and nonfacing sinuses.

The zones of apposition (commissures) are the areas on the coapting surface of the arterial valvar leaflets. Such zones extend to the middle of the valvular orifice and are not limited to the peripheral attachments of the free edge of the leaflets at the sinutubular junction.

The interleaflet triangles are the areas of arterial wall proximal to the attachments of the leaflets that are incorporated within the ventricular cavity in consequence of the semilunar nature of the attachments.

The ventriculoarterial junction is the zone of union between the muscular right ventricular infundibulum and the fibroelastic pulmonary arterial wall. When intact, it forms a circle or annulus. It is markedly discordant with the hemodynamic junction between the right ventricle and pulmonary trunk, the latter being produced by the semilunar attachment of the valvular leaflets (Fig. 1).

Results

Normal pulmonary root. As with the aortic valve, the pulmonary valve is a complex structure. Its most proximal part is the distal sleeve of the free-standing right ventricular infundibulum. This is muscular over its entire circumference, forming a cylindrical sleeve that can be removed from the right ventricle without encroaching on the cavity of the left ventricle. The pulmonary valvular leaflets, therefore, are at no point directly supported by the muscular ventricular septum. This sleeve of infundibular muscle gives rise to the fibroelastic wall of the pulmonary trunk at the anatomic ventriculoarterial junction (Fig. 1). This junction is seen as a straight line when the outflow tract is opened, but as a circle or ring when the root is intact. Inserted in the overall root are the leaflets of the pulmonary valve. They are attached in a semilunar fashion, with the locus of attachment marking the hemodynamic ventriculoarterial junction. This line of attachment crosses the anatomic ventriculoarterial junction at six points. Thus, the leaflets arise in part from the infundibular musculature and in part from the arterial wall. The semilunar line of attachment forms the border of the sinuses, with the sinus wall made up distally by arterial tissue and proximally by right ventricular musculature. Interposed proximally between the bases of the sinuses are the fibrous interleaflet triangles. Their borderlines are the line of attachment of the leaflets laterally and the right ventricular infundibulum basally. Their tips point toward the peripheral attachments of the zones of apposition between the leaflets. Distally the sinuses are confined by the sinutubular junction. This junction is not as pronounced as in the aortic root. A true ridge was found only in two (9%) of the normal
hearts obtained from children, and in two (22%) of the adult hearts. In all the hearts, nonetheless, it was possible to discern a clear line, separating the thin wall of the sinuses from the thicker wall of the pulmonary trunk, by transillumination through the translucent wall of the sinuses.

The leaflets were thickened along their semilunar locus of attachment between the sinutubular ridge and the muscular nadirs of the sinuses, analogous to the situation seen in the aortic valve (3). This coronet-like zone of attachment is more delicate than in the aortic root. Sagittal sections at various points along this semilunar line showed how the semilunar hinge of the leaflets was distinct from the fibrous anchorage of the arterial wall to the muscular infundibulum (Fig. 2). The fibrous accretion between the elastic arterial wall and the muscular infundibulum formed a seam at the anatomic ven-triculoarterial junction, in continuity with the fibrous interleaflet triangles and with the bases of the leaflets. The valvar leaflets were lined on their ventricular aspect by a thin layer of elastic tissue. The dense fibrous tissue on the arterial aspect was continuous with the fibrous accretion at their attachment.

Pulmonary valvular stenosis. It proved possible to distinguish between two groups of valvular stenosis, dome-shaped and dysplastic, with several less typical cases making up a third group (Table 1). The dome-shaped valves had a typical appearance. The valvular tissue in most cases was not thickened, but surrounded a central orifice in every case, the hole having a diameter of <1 mm in the neonatal or infant hearts. Three fibrous raphes originated at the level of the sinutubular junction and extended onto the surface of the valve toward the central orifice. These raphes supported the central orifice within the dome at the level of the sinutubular junction and, at the same time, tethered the arterial wall. The result was a waistlike narrowing of the sinutubular junction, but with basically normal arterial walls (Fig. 3). Because of this arrangement, the entrances to the sinuses were narrowed. The nadirs of the leaflets were, in most cases at, or only just below, the level of the ventriculoarterial junction. The base of the malformed valve was not attached in the same semilunar fashion as seen in the normal hearts. Instead the line of attachment of the domed valve was shallow, in some cases being almost circular (Fig. 3). Histologic analysis showed that the arterial wall was composed of well aligned layers of elastic tissue with smooth muscle and fibrous tissue in between, as in the normal pulmonary trunk. The sinutubular junction was more accentuated (Fig. 4), and the sinus wall was thinner than the truncal wall. In the case with combined dysplasia and doming of the valve, the proximal part of the leaflets was thin, but the distal part was considerably thickened and fibrous.

Table 1. Types of Stenosis

<table>
<thead>
<tr>
<th>Stenosis Type</th>
<th>No. of Patients</th>
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<tbody>
<tr>
<td>Dome-shaped valve, subtotal fusion of the zones of apposition, pinhole-sized orifice, leaflet tissue not thickened</td>
<td>8</td>
</tr>
<tr>
<td>Leaflets thickened, dysplastic leaflets, no fusion of the zones of apposition</td>
<td>6</td>
</tr>
<tr>
<td>Leaflets thickened, not dysplastic, partial fusion of the zones of apposition</td>
<td>2</td>
</tr>
<tr>
<td>One rudimentary leaflet, partial fusion of the zone of apposition</td>
<td>2</td>
</tr>
<tr>
<td>Bifoliate pulmonary valve</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>19</td>
</tr>
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The dysplastic valves showed cauliflower-like changes that severely thickened the leaflets and obstructed the right ventricular outflow merely by their size. The zones of apposition between the dysplastic leaflets were not fused, the sinuses were deep and the lines of attachment were semilunar, all as seen in normal hearts. In all cases only the distal parts of the leaflets were affected by the myxomatous degeneration; the most proximal part, originating from the right ventricular musculature, was smooth and delicate, but with a haphazard arrangement of fibrous tissue (Fig. 5).

The third group of hearts showed varying types of valvular stenosis, the specimens being unified by the presence of a degree of thickening of the leaflets, together with an asymmetric or partial fusion of the zones of apposition between them. Two hearts had a rudimentary right-facing leaflet and sinus, with incomplete fusion of the zone of apposition between the left- and nonfacing leaflets. This arrangement yielded a stenotic and eccentric orifice (Fig. 6a). The line of attachment was almost circular, with no true sinuses or interleaflet triangles seen. Two other hearts had three thickened leaflets of equal size, their zones of apposition having been opened by valvotomy. Again, the line of attachment was circular rather than semilunar, inhibiting the systolic opening of the valve even subsequent to the release of the leaflets. The final specimen had a bifoliate pulmonary valve, with leaflets of unequal size (Fig. 6b and c). The zone of apposition between the leaflets was oriented in a transverse direction, with the posterior, and larger, leaflet containing two shallow raphes. The line of attachment of the larger leaflet was semilunar and crossed the ventriculoarterial junction. The smaller leaflet, by contrast, was

Figure 3. a, Dome-shaped pulmonary valve seen from the arterial aspect. The raphes (v) draw the sinutubular junction toward the central pinhole orifice (arrow). b, Dome-shaped valve with deep sinuses (s) opened to show the plication of the arterial wall at the sinutubular junction. The arrow indicates the valvular orifice. c, Dome-shaped valve with shallow sinuses (s). The leaflets are slightly thickened at the edge and the line of attachment (broken line) is almost circular.

Figure 4. a, Dome-shaped valve with dysplastic leaflets. Three raphes (v) are seen. The arrow points to the central orifice. b, Histologic section shows fibrous dysplasia of the edge of the leaflets but a thinner component proximally. The broken line marks the boundary of the pulmonary infundibulum. The star indicates fibrous tissue between arterial wall and myocardium. c, The sinutubular junction (arrowhead) is markedly accentuated in this section taken close to the raphe. The fibrous membrane, analogous to the interleaflet triangle, is indicated by the star.

Figure 5. a, The peripheral attachments (arrowheads) of the dysplastic leaflets are not drawn inward. The sinutubular junction is not clearly demarcated. Although the leaflets are thickened in the distal parts, the proximal parts (arrows) are smooth and fairly thin. b, Histologic section through the depth of the sinus (s) shows the fibrous tissue (star) between the thin wall of the sinus and the myocardium.
shallow, with its proximal attachment at the level of the ventriculoarterial junction. The interleaflet triangles were of unequal size. The right one measured $10 \times 6$ mm, the left only $3 \times 4$ mm. Both leaflets were delicate, with no fusion of their zones of apposition, but their mobility was inhibited because of the abnormal architecture of the pulmonary root.

Discussion

Relatively little attention has been paid recently to the clinical anatomy of the pulmonary valve (2,7–12). This probably reflects, in part, the belief that the structure of the valve is well understood. It is also the case that in recent years the relief of stenosis has become the province of the interventional cardiologist rather than the surgeon, and the cardiologist views the abnormal substrate in the form of images rather than the real thing. It remains a fact, nonetheless, that those accounts that have appeared show startling discrepancies.

Clinical anatomy. As with descriptions of the aortic root, it is customary for surgeons, as well as pathologists, to describe the pulmonary “annulus” as a crucial structure in the pulmonary root (13). Gikonyo et al. (7), in their account on pulmonary stenosis, followed closely the description and the terminology used by Gross and Kugel in 1929 (14). They distinguished between supravalvular and subvalvular rings. After studying a substantial number of hearts, they illustrated their concept of the annulus by means of a single sagittal section taken at an unspecified location. Very recently, Hokken et al. (2) conceptualized a three-dimensional reconstruction of a collagenous valvar annulus based on serial sections taken in several planes. In our study we were not able to demonstrate any dissectable, clinically relevant, fibrous coronet in the pulmonary root. We fully appreciate the circular line of attachment and the nearly circular line of attachment and the large right interleaflet triangle. Zimmerman (11), in his report on valvular anatomy, emphasized how these fibers encircle the arterial root, but like a tricorn, rather than a ring. The fibers that reinforce the hinge are continuous with those that anchor the arterial wall itself to the muscular infundibulum. It is this second union that is a true circle but it, similarly, is not a discrete structure. Because the pulmonary root is subject to reduced hemodynamic forces compared with the aortic root, all these structures are more delicate and flexible than those seen in the normal aorta. Thus, the surgeon will not find a circular fibrous structure to remove or sew when dissecting the pulmonary root nor when inserting it in any other place. Instead, surgical attention should be focused on the free-standing muscular infundibulum. The free-standing nature of the infundibulum is often not well appreciated (15,16). In harvesting the pulmonary autograft in the Ross procedure, the risk of entering the left ventricular cavity, and of damaging the first septal artery, is increased if incisions are made inferior to the free-standing sleeve of musculature.

Substrates of valvar stenosis. Unless serial sections are taken across the entire pulmonary root, it is easy to be misled into assuming that a circular fibrous ring does, indeed, support the hinges of the leaflets. The tricorn arrangement becomes evident only when note is taken of the essential third (longitudinal) dimension of the semilunar line of attachment of the leaflet. This longitudinal extension, producing the semilunar attachments, is essential for the normal function of the valve, because it is this arrangement that gives the free edge of the leaflet the required degree of mobility for adequate diastolic closure and systolic opening. Thus, stenosis can occur in a semilunar valve that has an adequate width of the parietal structures of the outflow tract, even in valves with no fusion of the zones of apposition between the leaflets, simply because of a lack of “height” of the overall valvular apparatus. Such an arrangement was prominent in most of the dome-shaped pulmonary valves and in other nondysplastic, albeit stenotic, ones. Even when the zones of apposition are carefully liberated, such valves should not be expected to function normally. The leaflets may well open sufficiently, but at the cost of abnormal strain within them. It is tempting to argue that the typically dome-shaped valves are not themselves thickened...
because, due to extensive fusion and nonseparation of leaflets, they do not move at all.

As has been pointed out by Koretzky et al. (9), truly dysplastic pulmonary valves with secondary stenosis represent a different kind of abnormality from the valves with only fused zones of apposition. The cauliflower-like malformation of the distal part of the leaflet is most likely an intrinsic change and not due to a primary abnormal valvular architecture. The valves examined in our series showed all the necessary parietal features of a functioning semilunar valve. The sinuses, interleaflet triangles and the semilunar line of attachment of the leaflets were all of expected dimensions. The theory of an initially normal development of these valves is supported by our observations that the most proximal parts of the leaflets are not affected, or at least affected to much less a degree, by the underlying disease. We were unable to chart the clinical course of the patients from whom the hearts were obtained, so we cannot establish any relation to genetic or infectious disorders. From the clinical point of view, any successful dilation of such a valve is almost certainly precluded, because even adequate surgical debridement would result in nearly total removal of the leaflets. Milo et al. (17), in their account of pulmonary valvular stenosis, describe another type of obstruction produced by an hourglass-like narrowing and thickening of the pulmonary trunk at the level of the sinutubular junction. They emphasise the deep, bottle-shaped sinuses. Their description is reminiscent of the typical appearance of so-called supravalvular aortic stenosis. We did not find a similar specimen in our series, but our observations indicate that such a narrowing would represent a pathologic architecture of the complete valvular apparatus, and not a separate supravalvular phenomenon.

Conclusions. For adequate relief of pulmonary valvular stenosis, several surgeons have recommended partial or complete excision of the leaflets (17–20). This leads us to the ultimate question: do we need a pulmonary valve at all? Our investigation does not permit us to answer such a fundamental question. However, being aware of the excellent results of pulmonary valvoplasty, achieved either surgically or by balloon dilation, as well as of the good clinical state of many patients without a functioning pulmonary valve, we contend that our detailed analysis of a supposedly trivial thing, the morphology of pulmonary stenosis, has given valuable insight into more fundamental problems concerning the development, structure and function of the semilunar valves. Zimmerman (11) stated in 1966: “There is nothing uncomplicated about cardiac anatomy.” Is it more accurate to state that the complications are largely in the eyes of the beholders?

References