Review in Basic and Applied Anatomy

# Anatomic variations of the cardiac valves and papillary muscles of the right heart

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## Summary

This article reviews the right atrioventricular and pulmonary valves, along with their anatomic variations as well as the papillary muscles and chordae tendineae of the right ventricle of the human heart. A brief anatomical background is given for every structure, as well as a gross review of their embryological basis. Although the normal morphology of the right atrioventricular valve is tricuspid, this is not always the case; its anatomic variations involve, firstly, the number of cusps and accessory leaflets. Anatomic variations of the right atrioventricular valve may occur in association with other congenital anomalies and syndromes. Also the number, length and shape of the papillary muscles and chordae tendineae are variable. This can be of clinical significance since the papillary muscles play an important role in the contraction of the right ventricle and in the closure of the tricuspid valve so as to prevent ventricular blood from passing back into the right atrium. The pulmonary valve may present variations in the number of cusps, stenosis or atresia, either as isolated clinical findings or in association with congenital syndromes.

## Key to abbreviations
APM = anterior papillary muscle
AV = atrioventricular
LV = left ventricle
PPM = posterior papillary muscle
RV = right ventricle
SPM = septal papillary muscle
VSD = ventricular septum defect

## Introduction

The right ventricle (RV) of the human heart has 2 valves; the right, usually tricuspid, atrioventricular (AV) valve and the semilunar pulmonary valve. Papillary muscles which are located in the RV attach to the cusps of the AV valve via the chordae tendineae and contract to prevent inversion or prolapse of the valve. Together, the

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papillary muscles and the chordae tendineae are known as the subvalvular appara-
tus. There are three papillary muscles in the RV; the anterior (APM), posterior (PPM)
and septal (SPM) papillary muscles (Moore, 1992).

This review presents the most common anatomic variations of the heart valves
and the papillary muscles of the RV. A brief anatomical background is given for every
structure as well as a gross review of their developmental and embryological basis.

## Right atrioventricular (tricuspid) valve and papillary muscles

The right AV valve has three, roughly triangular shaped, cusps that project into
the ventricle: the anterior (superior), posterior (inferior) and septal. The anterior is the
largest cusp interposed between the AV orifice and the conus arteriosus. The poste-
rior is connected to the right margin of the ventricle and the septal cusp to the ven-
tricular septum. The bases of the cusps are attached to a fibrous ring at the AV orifice,
where they are continuous with one another. The margins of each cusp are irregularly
notched and thinner than the central portions. The ventricular surfaces of the cusps
are divided into three zones: (1) the distal rough zone, defined as the area between
the cusp's free edge and its line of closure, (2) the basal zone (only for the posterior
cusp) extending from the annulus of the posterior cusp to the clear zone and (3) the
proximal, thin and translucent clear zone which lies between the rough zone and the
basal one or the cusp base (Silver et al., 1971). The rough zone serves as points of
insertion for the chordae tendineae, which arise from the apices of conical muscular
projections of the ventricle wall, called papillary muscles (Moore, 1992; Walls, 1995).

The papillary muscles are the large APM (attached to the anterior wall of the
RV), with chordae inserted to the anterior and posterior cusps of the valve, the PPM,
often represented by two or more parts (attached to the inferior wall of the RV), with
chordae inserted to the posterior and septal cusps, and a variable group of small
septal papillary muscles (SPM), attached to the interventricular septum, with chor-
dae inserted to the anterior and septal cusps. According to a more recent functional
terminology for the tricuspid valve, the papillary muscles can be grouped according
to the distribution of their cords to a definite commissure and its contiguous main
leaflets. Therefore, the APM becomes the anteroposterior, the PPM the posteroseptal
and the SPM the anteroseptal papillary muscle, respectively (Joudinaud et al., 2006).
The papillary muscles contract just before contraction of the RV so as to tighten the
chordae tendineae and draw the cusps at the time ventricular contraction begins. This
prevents ventricular blood from passing back into the right atrium (Walls, 1995; Jou-
dinaud et al., 2006).

## Pulmonary valve

The pulmonary orifice is guarded by the pulmonary valve. The latter is composed
of three semilunar, pocket-like cusps (the anterior, right and left leaflet), whose con-
vex outer border is attached to the root of the pulmonary trunk. The free inner border
is thickened in the middle to form the nodule on each side of which there is a small,
thin crescent-like area (lunula). When the valve closes, following ventricular diastole,

the nodules and lunulae are pressed together projecting upwards into the lumen of the pulmonary trunk, thus preventing blood from returning into the RV (Walls, 1995; Joudinaud et al., 2006).

## Development of the tricuspid and pulmonary valves and papillary muscles

Beginning in the 5th week of gestation, three subendocardial tissue swellings located around the orifices of the pulmonary trunk will be the origins of the pulmonary valve as they will, eventually, hollow out and take the shape of three thin-walled cusps. The AV valve develops from the excavation of the supporting ventricular myocardium (Steding and Seidl, 1993). Cavitation of the ventricular walls forms a spongework of muscular bundles. Some of these remain as the trabeculae carneae and others become the papillary muscles and chordae tendineae which run from the papillary muscles to the AV valve (Moore and Perseaud, 1993; Mandarin-de-Lacerda, 1989). By the 8th week, the heart presents a complete AV valvular structure.

## Anatomic variations of the right AV valve and papillary muscles

Number of cusps and accessory leaflets

Although the right AV valve has three leaflets, autopsy series suggest that the number of leaflets may vary, or that accessory leaflets may be found between the main leaflets; like the former, they consist of a fold of endocardium strengthened by fibrous tissue (Walls, 1995). Even though researchers have tried to establish morphological and morphometrical criteria to distinguish between supernumerary and commissural cusps, no consensus has been achieved (Wafae et al., 1990). Commissural cusps are small accessory cusps occurring at the site of junction between adjacent cusps, i.e. at the site of the congenital fusion of the original commissures, that do not reach the fibrous ring of the valve (Fritsch, 2008). In a post mortem study, the right AV valve was not consistently tricuspid, but was observed to present with 2, 4, 5 or 6 cusps in 72% of cases; moreover, additional "commissural cusps" were found in 64% cases independent of the number of "supernumerary cusps" (Wafae et al., 1990).

In valves with 2 cusps, the septal cusp is larger than the anterior one. In valves with 3 cusps, the posterior cusp is associated with a less prominent septal cusp. When the valve comprises 4 cusps, the anterior and posterior ones become less prominent and an anterolateral cusp emerges. In this case, the posterior cusp, besides a reduction in size, undergoes medial dislocation assuming a more posterior position. In valves with 5 cusps, the posterior cusp does not change in size but undergoes lateral dislocation, assuming a position similar to that seen in valves with three cusps. In addition, the appearance of a posteromedial cusp leads to reduction in size of the septal one, while the anterior cusp undergoes little or no alteration. There is also some reduction in the size of the anterolateral cusp. When the valve consists of 6 cusps, the sixth one, named posterolateral, is located between the anterolateral and posterior cusps. Reduction in size is noted mainly in the septal and anterolateral cusps (Wafae et al., 1990).

An anatomic study of the AV valve in children showed that the commonest finding was 3 cusps, while a fourth cusp, if present, was classified as anterolateral in location. The number of tendinous cords was greater for the anterior and septal cusps than for the posterior and anterolateral cusps. In addition, the posterior region of the valve annulus in 35.7% of the cases was occupied by undeveloped valve tissue with the posterior valve located anteriorly in these cases (Gerola et al., 2001).

An accessory, mobile or fixed, right AV valve leaflet not contributing to the commissure is a rare finding, most often reported in children with complex congenital cardiac malformations (Fallot's tetralogy, Ebstein's anomaly). The mobile type is a parachute-like leaflet floating freely in the RV. The fixed type is firmly anchored to the interventricular septum by short chordae. When associated with ventricular septum defects (VSDs), both types can cause partial to near-complete obstruction of the VSD (Faggian et al., 1983; Hetzer et al., 1998; Yoshimura et al., 2000; Tewari and Mittal, 2006). Only a few cases are asymptomatic. In most cases, accessory valve tissue is associated with other cardiac malformations, therefore the symptoms of this anomaly depend on coexisting ones (Yoon et al., 2008). Accessory tissue originating from the tricuspid valve and protruding into the left ventricle (LV) outflow tract through a VSD has also been described. This tissue can form a pouch, the walls of which are similar to the tissue of the normal tricuspid valve, or can simply be a papillary-like mass of connective tissue. The end result can vary from mild to severe LV outflow tract obstruction. Adhesions to the rims of the VSD causing obstruction of the VSD can also be noted (Feigl et al, 1986).

## Double orifice right AV valve

Radermecker *et al*. (2011) reported on a double orifice right AV valve in a form of partial AV septal defect proposing leaflet fusion along part of their anticipated zones of apposition as an explanation for the formation of this anomaly. The tricuspid valve, divided into an anterior and a posterior orifice by a bridge of leaflet tissue, can cause valve stenosis because of the arcade deformity of the APM to which the bridging leaflet tissue has short chordal insertions, with the anterior orifice being regurgitant as a result of a deficient septal leaflet (Yoo et al., 1993).

## Tricuspid atresia

Tricuspid atresia refers to a congenital anomaly in which there is no physiologic or gross morphologic connection between the right atrium and RV and there is an interatrial connection allowing mixing of systemic and pulmonary venous return. There is a variable degree of hypoplasia of the RV. The LV and mitral valve may be normal (Gatzoulis et al., 2005). Prenatal diagnosis is possible through echography (Berg et al., 2010).

Valve atresia with complete absence of the three leaflets, chordae tendinae and papillary muscles, probably due to a primary pathogenetic step that occurs in the wall of the RV preventing the morphogenesis of the tricuspid valve, has also been reported with only the fibrous ring being present in the AV junction (Muñoz Castellanos et al., 1992).

Papillary muscles and chordae tendineae

The number, length and shape of papillary muscles and chordae tendineae in the RV are variable. This can be of clinical significance, since the papillary muscles play an important role in RV contraction by drawing the tricuspid annulus toward the apex, thereby causing shortening of the long axis and the chamber becoming spherical for ejecting blood (Hashimoto et al., 2001).

In one case series of 79 normal human hearts, the APM and PPM were present in 100% of the cases, while the SPM was absent in 21.5% of the hearts. The APM presented with 1 head in 81% of the cases and with 2 heads in 19% (Nigri et al., 2001). Double headed APM were, mostly, V or H shaped. In many cases, one large and one long APM were observed. Often, the APM presented with 3 or 4 papillae from which tendinous cords rose, heading to the anterior and posterior leaflets.

Regarding the PPM, 1 head was found in 25.4%, 2 heads in 46.8%, 3 heads in 21.5% and 4 heads in 6.3% cases. When more than one PPM was found, the average length was decreased so that the higher the number of papillae, the lower their length. The variability was even larger than for the SPM (one head in 41.7%, 2 heads in 16.5%, 3 heads in 12.7% and 4 heads in 7.6% cases).

The SPM was, in some cases, absent. When one SPM was present, it presented a small head with only one tip, giving insertion to tendinous cords heading to the septal and anterior leaflets. When more than one SPM was observed, each muscle had a very small head. The SPM may occur in two groups, one consisting of the constant musculus coni arteriosi and the second represented by other variable septal muscles. Jezyk *et al.* (2003) examined 111 human hearts; in the majority of them, the conus muscle mainly supplied the front leaflet, while, in the other septal muscles, provision of the front leaflet and the frontal part of the septal leaflet predominated. In 100 human hearts collected at autopsies, the conus muscle was present in 82% of the hearts; in the remaining 18%, it was replaced by tendinous cords. The papillary muscle of the conus was connected with the septal (59.7%), anterior (20.7%), or both septal and anterior leaflets (19.5%), through single (29.8%) or multiple chordae tendinae (70.1%). It was also found to be present as a single (51.8%), double (32.9%) or triple papilla (15.2%). Additionally, accessory single SPMs were identified in 42 specimens, double SPMs in 32 specimens and triple SPMs in 26 specimens (Loukas et al., 2009).

Since SPM displayed such wide morphological variations that its value as an anatomical landmark in the RV was very restricted, the name medial papillary complex was proposed. The medial papillary complex is represented by a main muscle belly, Lancisi's muscle, along with a variable number of adjacent minor papillary muscles and/or isolated cords. The chordae tendineae of Lancisi's muscle are attached to both the anterior and septal cusps of the tricuspid valve; sometimes insertion only to the anterior cusp is described (Wenink, 1977; Restivo et al., 1989). The mean number of chordae tendineae originating from the APM, PPM and SPM were 4.74, 2.67 and 1.77, respectively (Nigri et al., 2001).

Another study involving anatomic dissection of 400 human hearts reported that one-headed APM was found to be more frequent in patients that suffered from sudden cardiac death. Most of the PPMs were conical and flat topped. The results revealed an association between the absence of SPM, or lower ratio of APM to PPM, or both, with deaths of cardiac origin (Aktas et al., 2004).

The description of the chordae tendineae of the papillary muscles of the RV and their anatomic variations were reported by Silver *et al.* (1971) who depicted five different types of chordae tendineae attached to the tricuspid valve (Tab. 1). On average, 25 chordae were found to be inserted into the tricuspid valve, out of which 7 to the anterior leaflet, 6 to the posterior leaflet, 9 to the septal leaflet, and 3 into the commissural areas. Escande *et al.* (1980) described the mixed cord, one type of cylindric tendinous cord, always ramified either into two branches which are, in turn, greatly ramified or fan shaped. Each ramification is attached to the valve by an expansion which is either perpendicular or parallel to the ring. Other anatomists have introduced the concept of multi-apical and multi-segmental papillary muscles, with the former presenting as muscles which can be easily subdivided into more than one belly and the latter as muscles with more than one belly which cannot be easily subdivided from each other (Skwarek et al., 2005).

A study performed on a group of 107 formalin-fixed adult human hearts observed 3 types of connection between leaflets of the tricuspid valve and the papillary muscles: 1) a muscular connection (a straight connection between the leaflet and the papillary muscles), 2) a membranous connection and 3) the connection of tendinous cords in the tricuspid valve. Straight connections were found in 33 hearts studied (30.27%). In these hearts, a straight connection was present in one leaflet in 27 hearts (81.82%) and in 2 leaflets in 6 hearts (18.18%). Membranous connections between the tricuspid valve and papillary muscles were present in 7 of the 107 hearts studied (6.54%). The main leaflets usually received 20.79 ± 8.43 cords and the accessory cusps 8.14 ± 4.85 cords (Skwarek et al., 2006).

Aberrant tendinous cords, defined as those inserting to the clear zone of the tricuspid leaflet but not originating from papillary muscles, were retrospectively examined in 13.500 patients by echocardiography. Ten patients with aberrant tendinous cords tethering one or more tricuspid valve leaflets were identified. The presence of aberrant cords limiting the mobility of the tricuspid leaflets was identified as a cause of tricuspid regurgitation. There were short, non-aberrant tendinous cords in seven patients, five of whom also had RV or tricuspid annulus dilation (Kobza et al., 2004)

## The tricuspid valve in hearts affected by congenital anomalies and syndromes

The morphology of the tricuspid valve was analyzed in a total of 82 specimens with hypoplastic left heart syndrome that underwent post-mortem examination. A bicuspid right AV was revealed in 12% hearts; out of those, 15% had mitral stenosis and aortic atresia, and 38% combined stenosis. A quadricuspid AV was found in 2.4% of the cases. Accessory orifices were present in 2.4% hearts, all with mitral stenosis and aortic atresia. A moderately dysplastic tricuspid valve was observed in 33% specimens and a severely dysplastic tricuspid valve in 2%. The majority of the abnormalities were found in hearts with a patent mitral valve. The subvalvular apparatus was different in hearts with mitral atresia. A prominent APM was present in every heart examined, supporting the anterior leaflet as well as the zone of apposition between the anterior and the posterior leaflets. The PPM showed many variations of size and number of subunits. This was the case for all the abnormal hearts examined, irrespective of the patency of the mitral valve. Hypoplastic left heart syndrome represents a spectrum of congenital malformations, which, without surgical intervention, is

**Table 1** – Characteristics of the chordae tendineae of the papillary muscles of the right ventricle as described by Silver et al. (1971); the chordae are named according to their shape (the fan-shaped) or their site of insertion on the leaflet (the other types).

| Chordae tendineae | Zone of insertion | Insertion points | Characteristics |
|---|---|---|---|
| Fan-shaped | The commissures between the leaflets and into the clefts in the posterior leaflet | Anteroposterior commissure 94% Posteroseptal commissure 100% Anteroseptal commisure 82% | |
| Rough zone | The rough zone, into which most of the chordae tendineae are inserted, on the ventricular aspect of each leaflet. | Anterior leaflet 100% Posterior leaflet 82% Septal leaflet 98% | Each chorda splits into three cords soon after its origin One cord inserts to the free margin of the leaflet, one to the upper limit of the rough zone at the line of closure, and the third between the other two |
| Basal | A zone approximately 2 mm wide extending into the leaflet from the annular region | Anterior leaflet 46% Posterior leaflet 46% Septal leaflet 90% | Usually single and completely separated from the wall of the ventricle. They arise directly from the myocardium or from small trabeculae carneae and may flare into thin membranous bands just before their insertion. They may be round cords, flattened ribbon-like, long and slender, or short and muscular |
| Free edge | A leaflet free edge, frequently near its apex or else between the apex and a commissure or cleft. | Anterior leaflet 64% Posterior leaflet 48% Septal leaflet 50% | Single, threadlike, often long chordae usually originating from the apex of the papillary muscle but may come from its base. Sometimes they branch before insertion, and their fine subdivisions form a delta-shaped insertion at the free edge. |
| Deep | Leaflets ventricular surface either in the upper part of the rough zone or in the clear zone | Anterior leaflet 76% posterior leaflet 58% Septal leaflet 66% Anteroposterior commissural area 2% | They may be single or they may branch into two or three cords just before insertion |

always fatal within the first weeks of life; the features described here should be considered in reconstructive operations (Stamm et al., 1997).

Even more rare anatomic variants include: 1) duplication of the tricuspid valve, 2) an abnormal attachment of the tricuspid septal leaflet to a PPM associated with fusion of the right ventricular SPM and APM, and 3) a fenestration defect of the septal leaflet of the tricuspid valve. The prognosis of these variants is poor, as they are associated with other congenital anomalies such as Ebstein's anomaly or ectodermal dysplasia (Miyamura et al., 1984; Prayson et al., 1990). Absent pulmonary valve associated with membranous tricuspid atresia or severe tricuspid stenosis, intact ventricular septum and patent ductus arteriosus have been reported sporadically in the literature. Early diagnosis during pregnancy is associated with favourable prognosis after palliative treatment (Lato et al., 2010). Extreme cases of imperforate tricuspid valve, sometimes associated with dysplasia of the pulmonary valve, have also been described (Mori et al., 1992). The congenitally unguarded orifice is a rare lesion in which the tricuspid orifice is completely devoid of leaflet tissue (associated with pulmonary atresia and intact ventricular septum) (Anderson *et al.*, 1990).

Congenital tricuspid incompetence due to valvular dysplasia is a defect involving the leaflets (normally inserted on the ring), the chordae tendinae and papillary muscles of the tricuspid valve. This condition is rare, usually diagnosed during surgery. Tricuspid valvular dysplasia was defined by Becker *et al.* (1971) to include the spectrum of lesions involving a) focal or diffuse thickening of the valve leaflets, b) deficient development of cords and papillary muscles, c) improper separation of valve components from the ventricular wall and d) focal agenesia of valvular tissue. Congenital tricuspid valve dysplasia is categorized into grades; the grade is estimated from the most severe alteration present anywhere in the valve. There are three grades (minimal, intermediate and severe) according to what is found for the valve leaflets, the cords and the subvalvular apparatus (Tab. 2). Recent cases have demonstrated distinct forms of congenital tricuspid valve anomaly in which tricuspid regurgitation is due to short tendinous cords tethering the septal leaflet and preventing coaptation (McElhinney et al., 1999).

Another morphologic feature which causes malformation of the tricuspid valve is Ebstein's anomaly. In this rare and complex disorder, the primary lesion is downward displacement of the basal attachment of the posterior and septal leaflets (Lang et al., 1991). Ebstein's malformation is, in essence, a manifestation of abnormal development of both the myocardium and the valve components. During development of the normal heart, the leaflet tissue of the valves evolves from the supporting ventricular myocardium in a process that has been termed "delamination".

Ebstein's anomaly is characterized by adhesion of the septal and posterior tricuspid valve leaflets to the underlying myocardium due to failure of delamination. This leads to apical displacement of the annulus of the tricuspid valve. The attachment of the skirt of leaflet tissue guarding the entrance to the RV is not to the AV junction but rather to the wall of the ventricle itself (Martinez et al., 2006). The anterior leaflet may be severely deformed and may form a large 'sail-like' intracavitary curtain, which can even lead to RV outflow tract obstruction (Alonso-Gonzalez et al., 2010).

The straddling tricuspid valve, defined as the biventricular insertion of the tensor apparatus (chordae tendineae and papillary muscles) of the tricuspid valve, remains an incompletely understood form of complex congenital heart disease. Misalignment

Table 2 – Grades of congenital tricuspid valvular dysplasia described by Becker et al. (1971)

| Grade | Characteristics | | |
|-------|-----------------|--|--|
| | **Valvular Leaflets** | **Chordae Tendineae** | **Subvalvular Apparatus** |
| Minimal | nodular and thickened | normal | normal |
| Intermediate | thickened, longer than normal, often irregularly shaped | fibrous and thickened, short, abnormally attached to leaflets | underdeveloped papillary muscles, direct attachment of leaflets to papillary muscles or unspecialized parts of the right ventricle, but with still wide spaces between leaflets and ventricular wall |
| Severe | thickened, elongated, with irregular fenestrations, areas of focal agenesis (including agenesis of entire leaflets) | rudimentary or absent | abnormally close adhesion of valvular tissue to the right ventricle wall, with broad areas of leaflets approaching the ventricular wall with little or no space between the leaflet and the wall and varying amounts of myocardial tissue on the ventricular aspect of attached valve leaflets |

of atrial and ventricular septa is an essential feature of a straddling tricuspid valve, creating an inlet septal defect. Across this defect, the tricuspid valve straddles into the LV, where it is separated from the mitral valve by a posterior muscular ridge, the posteromedial muscle (Wenink and Gittenberger-de Groot, 1982). A morphometric study of 19 post-mortem cases of straddling tricuspid valve was performed with the results being compared with 32 normal control heart specimens. In straddling tricuspid valve, marked misalignment of the ventricles was always found relative to the atria. The normal angle between the ventricular septum and the atrial septum in the short-axis projection averaged 5°±2°. In contrast, in the 19 hearts examined with straddling tricuspid valve, the ventriculoatrial septal angle averaged 61°±24°. A VSD was present in 79% cases, AV canal type in 42%, AV canal type confluent with a conoventricular defect in 26%, and a conoventricular defect in 11% (Pessotto et al., 1999).

Double inlet left ventricle is a congenital heart defect that affects the valves and chambers of the heart. The RV is frequently small and both the mitral and tricuspid valves open into the enlarged LV, which is on the right-hand side of the body. In addition, there are defects in both the atrial and ventricular septa. Sixty-three autopsied hearts with double inlet LV and isomeric atrial appendages were studied; a valve with three or four leaflets was seen more frequently in hearts with double inlet LV. Complicated multiple orifices within the valve curtain, including abnormal accessory orifices within a leaflet, were found in 7 cases with double inlet to a dominant LV or RV. The presence of four papillary muscles was the most common pattern in hearts with double inlet LV. Straddling of the papillary muscles to a rudimentary and

incomplete ventricle was seen in 23% of cases. Direct attachment of tendinous cords to the ventricular septum or parietal wall was seen in 81% hearts with double inlet (Uemura et al., 1998).

## Anatomic variations of the pulmonary valve

### Number of cusps

Bicuspid pulmonary valve is a rare congenital disorder usually associated with other congenital heart defects. In a reported case of bicuspid pulmonary valve, imaging studies revealed a valve with anterior and posterior commissures and a mildly restricted opening (Vedanthan et al., 2009). Another rare congenital anomaly which is almost always discovered in post-mortem studies is quadricuspid pulmonary valve. Usually, it is an isolated anomaly with imaging studies confirming four cusps of similar size with a non-significant pulmonary insufficiency and a great aneurysm of the pulmonary artery (Gentille Lorente, 2009).

Bicuspid and quadricuspid pulmonary valves are, usually, considered as minor cardiac defects because of their mild clinical relevance (Roberts, 1993). The abnormal architecture of the pulmonary valve rarely alters the function of the valve itself and the anomaly often remains silent (Ricci et al., 2005). Quadricuspid pulmonary valve has also been reported in an infant patient with misalignment of pulmonary vessels and alveolar capillary dysplasia (Roth et al., 2006).

A case of reported pentacuspid pulmonary valve showed the valve with three almost equal cusps and two smaller cusps. All cardiac valves had a normal anatomic structure, and no clinical or biological evidence of heart failure was found (Demircin and Keles-Coskun, 2010).

### Pulmonary stenosis

Congenital pulmonary valve stenosis with intact ventricular septum is by far the commonest cause of obstruction to the RV outflow and is a relatively common lesion. Supravalvular pulmonary stenosis can also lead to RV outflow obstruction. In isolated pulmonary stenosis, the valve is usually acommissural unicuspid with a central stenotic orifice (Roberts et al., 1973; Polansky *et al.*, 1985; Freedom, 1983). The anatomy of tetralogy of Fallot with pulmonary stenosis was studied by Anderson and Jacobs (2008). A narrowing of the RV outflow tract occurred at the pulmonary valve (valvular stenosis) or just below the pulmonary valve (infundibular stenosis). The later was produced by the 'squeeze' between the anterocephalad misalignment of the outlet septum and the abnormal situated hypertrophied septoparietal trabeculations which extend onto the ventricular free wall. Cases were also found of stenosis more proximal within the ventricle, produced either by hypertrophy of the moderator band or by prominent apical trabeculations, giving a 'two-chambered RV'.

Abnormal pulmonary valves may be classified as acommissural with a prominent systolic doming of the valve cusps and an eccentric orifice, unicommissural with a single asymmetric commissure, bicuspid with fused commissures, or dysplastic with severely thickened and deformed valve cusps (Kirshenbaum, 1987).

A distinctive type of pulmonary valvular stenosis, termed "pulmonary valvular dysplasia", is a deformity characterized by the presence of three distinct cusps and commissures and composed of disorganized myxomatous tissue with little, if any, fusion. The obstructive mechanism is related to the markedly thickened, immobile cusps (Koretzky et al., 1969).

Pulmonary atresia

The extreme of pulmonary valve stenosis is pulmonary atresia with or without VSD. Cases of pulmonary atresia have been reported, usually accompanied by some other cardiac abnormality like tricuspid atresia or dysplasia of the RV free wall (Kasahara et al., 2010; L'Ecuyer et al., 2001).

The pulmonary valve in hearts affected by congenital anomalies and syndromes

Ten percent of individuals with Fallot's tetralogy have pulmonary atresia rather than pulmonary stenosis. In addition to the unrestricted VSD and anterocephalad deviation of the outlet septum, there is an absence of direct communication between the RV cavity and the pulmonary trunk. This lack of communication can be subvalvular/muscular or valvular. This lesion has also been referred to as "pulmonary atresia with VSD". Pulmonary atresia with intact ventricular septum is a disorder that involves the whole RV. An associated Ebstein's deformity of the tricuspid valve is found in 10% of the cases, further complicating the anatomy and the function of the RV (Stellin et al., 1993). The absent pulmonary valve syndrome is a rare cardiac malformation, usually associated with Fallot's tetralogy. It is usually associated with a VSD, stenotic pulmonary annulus, and RV hypertrophy (Stafford et al., 1973). Other cardiac anomalies associated with absent pulmonary valve syndrome include atrial septal defect, right aortic arch, patent ductus arteriosus and left pulmonary artery arising from the ductus arteriosus (Godart et al., 1996; Wu, 2008). Congenital absence of the leaflets of the pulmonary valve is less common when the ventricular septum is intact. Characteristic features of the syndrome include dysplasia or absence of the pulmonary valve leaflets permitting severe pulmonary regurgitation and aneurysmal dilation of the pulmonary arteries (Zucker et al., 2004; Waller et al., 1995). The combination of absent pulmonary valve syndrome and absent left pulmonary artery has also been described, causing higher morbidity and mortality (Abbag, 2006). Absent pulmonary valve syndrome has been reported with aneurysmal dilation of the ascending aorta (Liang et al., 2010). A combination of absent pulmonary valve along with malformed tricuspid valve and a dysplastic RV is a very rare syndrome (Nakamura et al., 2010).

Pulmonary stenosis (caused by dysplasia of the pulmonary valve) can be encountered in patients with multiple malformations such as Noonan, LEOPARD (multiple lentigines, electrocardiographic-conduction abnormalities, ocular hypertelorism, pulmonary stenosis, abnormal genitalia, retardation of growth, and sensorineural deafness) or Watson syndromes (Mendez and Opitz, 1985; Musewe et al., 1987; Loukas et al., 2004).

Pulmonary trunk dilation is reported in association with pulmonary valve abnormalities or with bicuspid aortic valve in the absence of pulmonary valve abnormality,

suggesting primary vessel wall pathology predisposing to arterial dilation. A systemic abnormality of connective tissue common to both arteries may be responsible, or dilation may result from a common developmental exposure as both originate from the embryologic cono-truncus (Kutty et al., 2010).

## Conclusion

Although the normal morphology of the right AV is tricuspid this is not always the case; its anatomic variations involve, firstly, the number of cusps and accessory leaflets. Two up to six cusps have been reported (most of the times discovered incidentally, postmortem, during autopsy). Accessory tricuspid valve leaflets, although rare, are most often reported in children with complex congenital cardiac malformations, rendering their management by the clinician complex, as other problems need to be clinically addressed, like LV outflow tract obstruction and VSDs.

Cases of double orifice right AV valve have also been reported, in which the surgeon is faced with the problem of valve stenosis. Tricuspid atresia is associated with a variable degree of hypoplasia of the RV. Finally, the anatomy of the tricuspid valve is somewhat altered in patients with congenital anomalies (not only of the heart); hypoplastic left heart syndrome requires reconstructive operations of a number of congenital cardiac malformations (mitral stenosis, aortic atresia, dysplastic tricuspid valve). Extreme cases of imperforate tricuspid valve or of a congenitally unguarded orifice have also been described. Tricuspid valvular dysplasia (ranging from altered anatomy of the valve itself to deficient development of cords and papillary muscles) can be a challenge for the surgeon depending on its severity and grade.

The number, length and shape of the papillary muscles and chordae tendineae are variable. This can be of clinical significance since the papillary muscles play an important role in RV contraction and in the closure of the tricuspid valve so as to prevent ventricular blood from passing back into the right atrium.

Also, for the pulmonary valve, the number of cusps is variable. Except from its normal tricuspid morphology, 2 to 5 cusps have been described (usually with no clinical significance). Congenital pulmonary valve stenosis is the most common cause of obstruction to the RV outflow, with pulmonary atresia representing the worst case scenario (with or without VSD). Pulmonary atresia (or even absent pulmonary valve) can be found in patients with congenital cardiac anomalies (e.g. Fallot's tetralogy), associated with VSD and absence of direct communication between the RV and the pulmonary trunk.

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