CONGENITAL LESIONS OF THE TRICUSPID VALVE

Review of Literature
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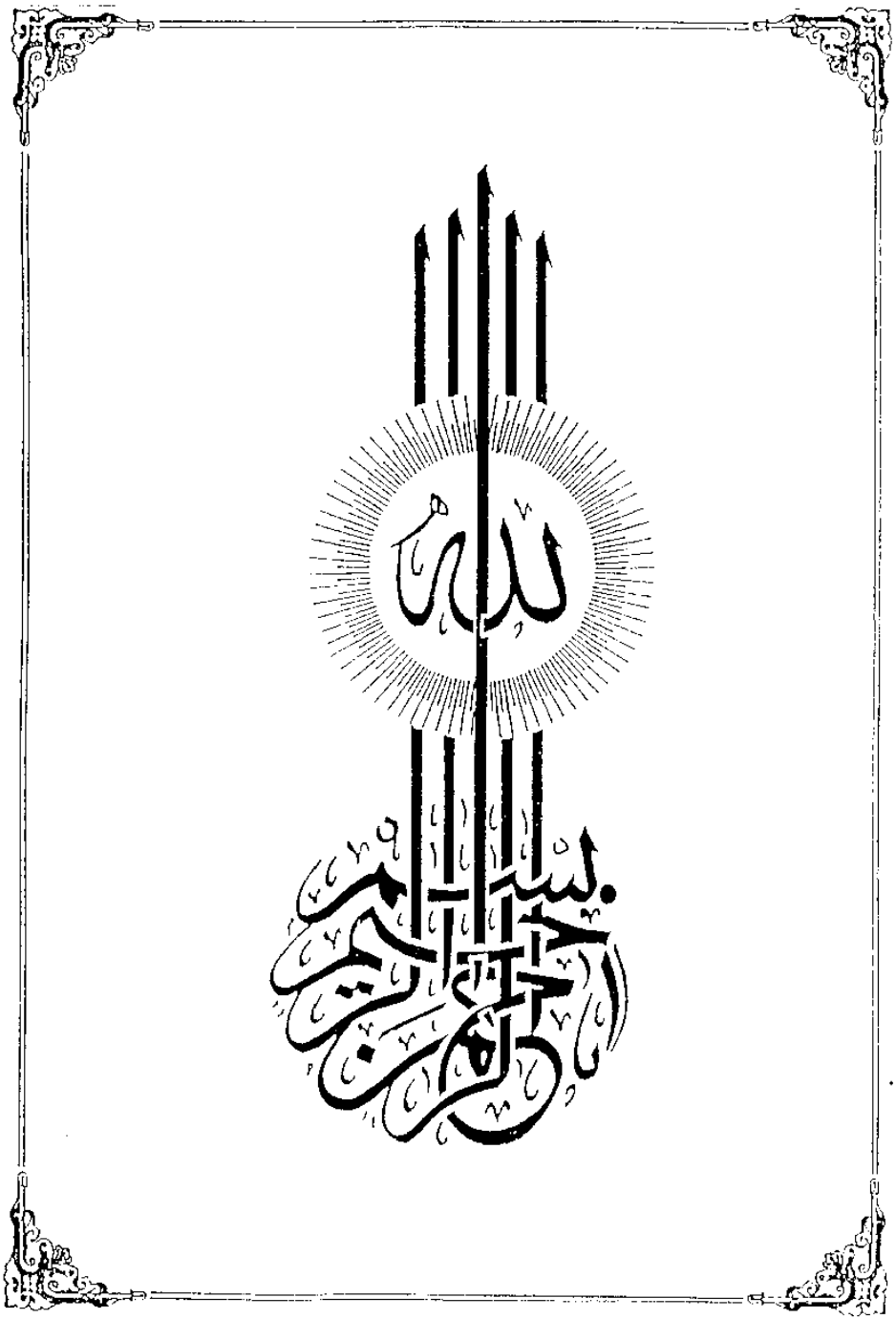

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INTRODUCTION

There is a wide vareity of congenital lesions which may affect the tricuspid valve.

These congenital lesions are :

1. Tricuspid atresia.
2. Ebstein's anomaly of the tricuspid valve.
3. Congenital tricuspid stenosis.
4. Congenital tricuspid incompetence.
5. Tricuspid valve prolapse.
6. Straddling of the tricuspid valve.

Embryology of the tricuspid valve

During expansion of the atrioventricular canal the cardiac jelly becomes cellular and is molded into two major cushions superior and inferior and two smaller lateral atrioventricular cushions. The superior cushion is attached to the roof of the atrioventricular canal while the inferior cushion lies on its floor.

As a result of protrusion of the cushions into the lumen of the A.V. Canal, the flow is divided into two streams, mitral and tricuspid.

The ventricular aspect of these two cushions is concave, so that their lateral ends protrude into the ventricle like horns known as the tubercles, hence there are two right and two left tubercles, superior and inferior on each side. Van Mierop L.H.S. et al., 1962.

Fusion of the superior and inferior atrioventricular cushions results in the formation of the septum of the canal. This septum divides the canal into the mitral and tricuspid channels. Gray's, 1973.

The lateral cushions are initially small, but later in their development they become somewhat more prominent.

Invagination of the A.V. cushions into the right ventricle (the tricuspid flange) contributes in the formation of the anterior and posterior leaflets of the tricuspid valve. Goor et al., 1970.

The endocardial cushion tissue also shares in the development of the septal leaflet of the tricuspid valve. Friedman, 1984.

However, the formation of the definitive A.V. valves which starts when the embryo is about 12 mm. C.R. length (Crown to rump) is derived in only very small part from the endocardial cushion tissue while the remainder of the material is derived from the muscular ventricular wall, the internal layer of which is liberated by a process of diverticulation and undermining. A skirit of ventricular muscle is formed at each A-V. orifice originating from the A-V. junction and attached lower down to the ventricular walls or septum by trabecula retained for this purpose. Van Mierop L.H.S. 1969.

All A.V. valves are therefore thick and fleshy at first, and only later in development, they are transformed into thin and fibrous cusps. Van Mierop L.H.S., et al. 1962 .

The chordae tendinae like the valve cusps are initially thick, muscular and few in number only later they are transformed into delicate fibrous strands. The papillary muscles remain muscular. Van Mierop L.H.S. and Kutsche, 1982.

The anterior cusp of the tricuspid valve is the first to develop and when the embryo is about 15-16 mm. C.R. Length, the anterior cusp is free from the ventricular wall. This is probably the reason that it's origin is normal in cases of Ebstein's anomaly of the tricuspid valve.

The posterior leaflet is formed after the anterior one and finally the septal leaflet is formed which in the adult overlies the membranous septum. The septal leaflet is formed when the embryo is about 50 mm. C.R. length (10-12 week old fetus). Van Mierop L.H.S. 1969.

Tricuspid Atresia

Definition

Tricuspid atresia as a matter of fact belongs to the group of cardiac lesions causing cyanosis, it is characterized by obstruction of the tricuspid orifice resulting in a lack of communication between the right atrium and right ventricle.

Three other defects are always associated with this condition (Peter Vlad, 1978).

1. Patency of the atrial septum.
2. Enlargement of the mitral valve and left ventricle.
3. Hypoplasia or absence of the right ventricle.

Prevalence

Tricuspid atresia is the thrid most common cyanotic lesion following Fallot's tetralogy and transposition of the great vessels.

It occurs in about 3% of postmortem and clinical congenital heart disease,(Peter Vlad, 1978).

Embryology

Van Praagh, 1973 has postulated that tricuspid atresia is due to malaligment of the ventricular septum with the atrioventricular canal. Absence of the right ventricular sinus results in shifting of the ventricular septum to the right and obliteration of the right atrio ventricular orifice.

Tricuspid atresia can thus occur with complete atrioventricular canal.

Rosenquist et al., 1970 observed that the right atrial dimple which is the presumed site of the atretic valve transilluminates into the left ventricle regardeless of the anatomic type, size of the ventricular septum or anatomy of the atrial septum.

Anatomy of Tricuspid Atresia

The common anatomical features of tricupsid atresia are.

A. Atresia of the tricuspid orifice

There is agenesis of the orifice, there is no connection between the right atrium and right ventricle. Scalia et al., 1984.

Weinberg 1980, studied thrity three autopsy cases of tricuspia atresia and described five distinct types :

1. Muscular Type :

It occurs in 76% of the cases, in which the right atrium has a muscular floor, sometimes a tiny dimple is present in the expected site of the tricuspid valve.

2. Membranous type :

It is present in 12% of the cases, in this type the atrio ventricular portion of the membranous septum between the right atrium and left ventricle appears in the expected location of the tricuspid valve.

3. Valvular type :

It occurs in 6% of the cases, in which there is a thin membrane of valvular tissue sometimes with radimentary chordae, there is no atrioventricular orifice, just a solid mass of fused leaflets which is present between the right ventricle and the right atrium.

4. Ebstein's type :

It occurs also in 6% of the cases, in which the leaflet tissue is adherent to the wall of the already small right ventricle further reducing it's size.

5. Common atrioventricular canal type :

In this type a leaflet of the common atriventricular canal completely seals the only entrance into the right ventricle. However this type is very rare.

B. Interatrial communication :

There must be an atrial septal defect of any type in order to provide an exit of blood from the right atrium into the left atrium. It is usually a patent foramen ovale, it is present in two thrids of the group studied by Peter Vlad. 1978, which is 143 cases or a secundum atrial septal defect.

The size of the patency varies markedly depending chiefly on the age. It may be slit like in infancy and larger in older age.

A restrictive atrial communication in a sense that the diameter of the defect is less than that of the aorta in right sided tricuspid

valve and the pulmonary artery in left sided valve is uncommon in patients with normally related great vessels but occurs in about half the cases with d-transposition and all cases with L-transposition. Weinberg, 1980.

Aneurysm of the fossa ovale may occur in association with this type of restrictive atrial communication, when this aneurysm is large, it may cause mitral valve obstruction. Freedom and Rowe 1978.

Occasionally the aneurysm of the atrial septum may cause obstruction of the pulmonary veins leading to acute or chronic pulmonary congestion. This may be identified by echocardiography and proved by cardiac cathelerization and angiocardiography and it is of special interest to the surgeon. Reder et al., 1981.

Usually the interatrial comunication allows an obligatory right to left shunting of blood. Perloff, 1978.

However, Rao 1983, observed the presence of a physiological left to right shunting of blood at atrial level in 27 cases out of 48 cases studied. This is proved by the presence of a higher right atrial oxygen saturation than the superior vena caval saturation. After excluding the anatomical causes of a left to right atrial shunts such as ostium primum defect, total anomalous pulmonary venous drainage, ruptured sinus of valsalva into the right atrium and ruptured coronary sinus into the right atrium, thus the higher right atrial saturation is due to a physiological left to right shunt.

Rao 1983, also observed that the left to right shunt occurs during diastole as evidenced by the higher left atrial diastolic pressure than the right atrial diastolic pressure while in systole the right atrial pressure is higher, hence the shunt is right to left in systole.

However this left to right shunt is of no concern and it may be due to the increased pulmonary blood flow leading to left atrial enlargement and stretching of the foramen ovale. Rao 1983.

C. Mitral valve and left ventricle:

In tricuspid atresia, the mitral valve through which the systemic and pulmonary venous return must pass is larger than normal but of normal shape and competent. Laks H. 1983.

The mitral valve may be overriding the ventricular septum. Fragoyannis and Kardalinos 1962. The left ventricle is also enlarged and hypertrophied as expected from the increased work load. Laks H. 1983.

D. The right ventricle :

In some cases the right ventricle is absent or may be a slit like space or a microscopic finding within the wall of the left ventricle beneath the pulmonary valve and artery.

However, there is usually a small right ventricular cavity several millimeters in diameter formed of two compartments,

a trabeculated lower portion extending to the acute margin of the heart with rudimentary papillary muscles, the small ventricular septal defect usually communicates with the upper part of this portion, above this there is a small infundibular portion which may present an obstruction beneath the pulmonary valve and is continuous with the pulmonary artery. Peter Vlad, 1978.

In other cases the right ventricular cavity is larger and communicates with the left ventricle through a large ventricular septal defect . Laks H., 1983.

Fragoyannis and Kardalinos 1962 reported a case in which the right ventricle was the major chamber with the mitral valve overriding the septum.

Peter Vlad, 1978 described a case in which there was ventricular inversion, with a left sided dominant right ventricle and a hypoplastic right sided left ventricle.

Deanfield et al., 1982 in their study of 48 cases of tricuspid atresia found that the inlet portion of the right ventricle is absent in all and that the right ventricle is separated from the left ventricle by a septum which does not extend to the crux of the heart which is the site of meeting between the right and left atrioventricular grooves and the posterior interventricular septum, also this septum is grossly malaligned relative to the atrial septum.

The size of the right ventricle ordinarily correlates with the size of the ventricular septal defect. Laks H., 1983.