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"Unusual drainage of a single vertical vein in TAPVC in a case of Heterotaxy syndrome (right atrial isomerism) – auniqueanatomical variant"

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ABSTRACT: A rare type of Total anomalous pulmonary venous connection (TAPVC) in which a single vertical vein from common pulmonary venous chamber is draining in to two different systemic venous systems, one in to superior vena cava through innominate and other into inferior vena cava through hemiazygous.

I. INTRODUCTION:

Heterotaxy is a rare malformation syndrome with incidence of 1-1.5/10,000 live births [1-3]. These are associated with wide range of complex cardiac and venous anomalies[4].Heterotaxy syndrome especially right atrial isomerismis frequently associated with anomalies of pulmonary venous drainage[5].

Total anomalous pulmonary venous connections are classified based on the site of abnormal connections by Craig,Darling and Rothney[6].TheMixed TAPVC are categorised by Chowdhury UK et.al which is also based on anatomical site of abnormal connections[7].

TAPVC associated with heterotaxy syndrome will have a bizarreanatomicalconnections[7], which are very difficult to categorize by using present classification systems.

We are reportinga rare case of TAPVC that does not fit in to any classification system.

CLINICAL DETAILS:

A 6yrs old girl was referred for evaluation of complex cyanotic congenital heart disease.At the time of presentation,she had saturation of about 70% on room air.

An echocardiogram suggested diagnosis of situs ambiguous right atrial isomerism with complete AV canal defect with L-looping of ventricles, DORV,TGA VSD and PS.All pulmonary veins were forming a common venous chamber and opening into innominate through a vertical vein.

Cardiac CT was done to confirm echo findings and surgical planning. It was decided to go ahead with single ventricle palliation(figure.1).

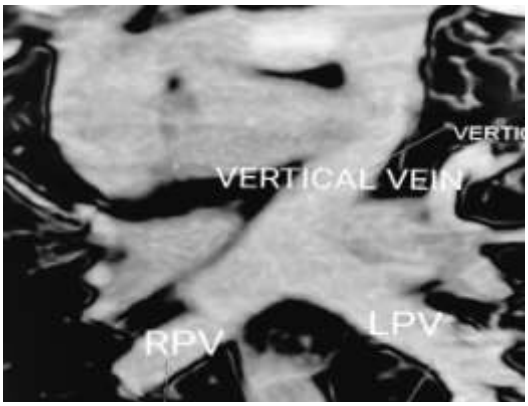

Figure-1: -CT-angiography-Supra cardiac TAPVC

Diagnostic angiography was planned to confirm operability. On selective injection in the vertical vein, incidentally an additional vein was demonstrated,originating from the vertical vein, coursing along the spine and draining in to IVC system (Hemiazygos). This vein was missed in CT contrast study, because of non-opacification, in absence of aselective injection. Azygos vein was absent(figure.2)

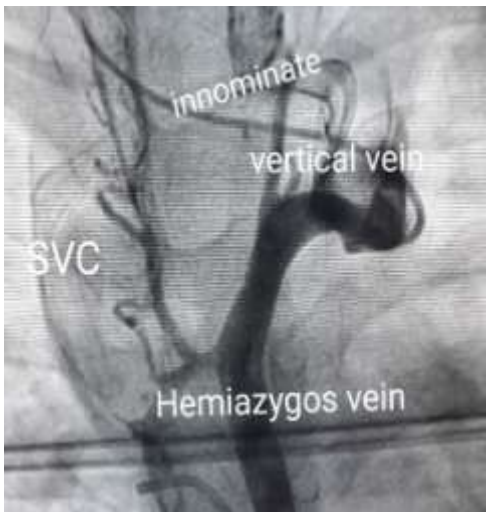

Figure-2: -Selective angiography of hemiazygos vein

Finally, surgical plan was made to do Bi-Directional Glenn shunt with TAPVC repair along

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with ligation of vertical vein and additional vein joining with vertical vein(Hemiazygos vein).

Intraoperatively, preoperativefindings were confirmed.All the four pulmonary veins were joined to form a common venous chamber and opened in to a vertical vein.The vertical vein which is posterior to LPA joined with innominate vein.An additional vein which was found incidentally during angiography was identified.It joined with vertical vein just above the left upper pulmonary vein(fig.3,4&5).

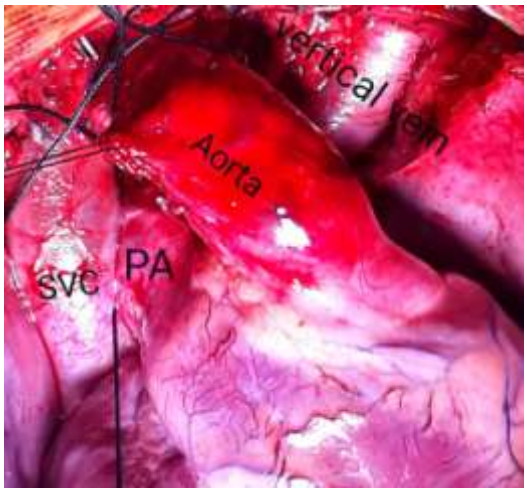

Figure-3: -Relation of aorta, pulmonary trunk, SVC and vertical vein

Figure-4: -Vertical vein passing posterior to LPA

Figure-5: -Hemiazygos joining with vertical vein just below the LPA

TAPVC repairwas done using posterior approach with ligation and division of vertical vein as well as the additional vein along with Bi Directional Glenn shunt.

Postoperative course in the hospital was uneventful and discharged in a satisfactory condition.

II. DISCUSSION:

Heterotaxy syndromes are associated with anomalies of pulmonary venous and systemic venous systems[8]. The TAPVC that are associated with heterotaxy syndromes have bizarre anatomy[7],which will be difficult to categorize with present existing classification.

Recently cases of such an unusualanatomical variant of TAPVC have been reported[9,10,11]

To the best of our knowledge,the anatomical variant of TAPVC, seen in our case, has not been reported previously.

In our case there was a single vertical vein which was draining into innominate vein as in usual supracardiac TAPVC,but during diagnostic angiographic study we found incidentally an additional vein (hemiazygos) joined with vertical vein. It was missed in the CT contrast study. Thus, vertical vein had two communications one with superior vena cava through the innominate vein and other with inferior vena cava through the hemiazygos.

There was a possibility that, this unusual second communications with systemic venous system,may remain patent after initial surgery due to increased pressure in common venous chamber and a constant source of persistent abnormal pulmonary venous drainage to systemic venous system[10].

The fate of such, unligated vertical vein or missed vertical vein lead to persistent left to right shunting and volume overload on right side of the heart[12,13,14,15].

In the case we reported,additional vein was ligated and divided since italso causing distortion of anastomosis near the left upper pulmonary vein.

Hence in heterotaxy syndromes for detail study of venous anomalies, different modalities of investigations are required for better outcomes.

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