Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Complete Abnormal Venous Return Associated with Complex Heart Disease: Case Report

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DOI: 10.36347/sjmcr.2024.v12i04.035 | **Received:** 05.02.2023 | **Accepted:** 12.03.2024 | **Published:** 27.04.2024

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Abstract **Case Report**

Total anomalous pulmonary venous return (TAPVR) is a cardiac malformation defined by a complete absence of communication between the pulmonary veins and the left atrium. All oxygenated pulmonary venous return flows directly or indirectly into the right atrium. Survival is only possible via a right-to-left atrial shunt. Non-blocked forms present as high-flow shunts with moderate cyanosis. Symptoms in blocked forms are dominated by obstruction of venous return, and life-threatening distress with intense cyanosis and severe pulmonary hypertension sets in from the very first days of life. Treatment consists in connecting the pulmonary venous receptacle to the left atrium, tying off the collector and closing the inter-atrial septum. The risks of surgery remain high in severe forms, and failures usually occur in the first year of life. In the case of good surgical correction, the long-term prognosis is excellent.

Keywords: pulmonary veins and the left atrium, (TAPVR), atrial shunt, prognosis.

INTRODUCTION

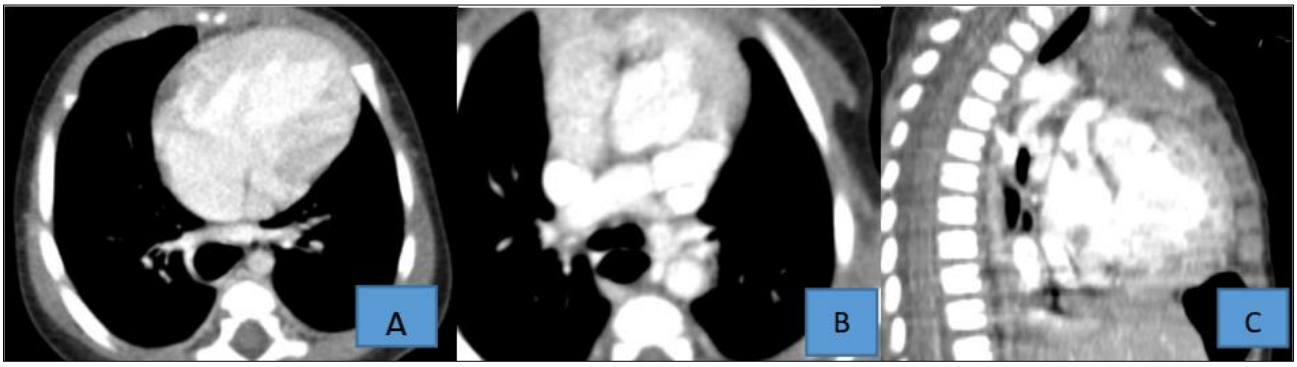
Pulmonary venous return anomalies include pulmonary vein connection anomalies (total or partial anomalous pulmonary venous return), and congenital atresia or stenosis of one or more veins.

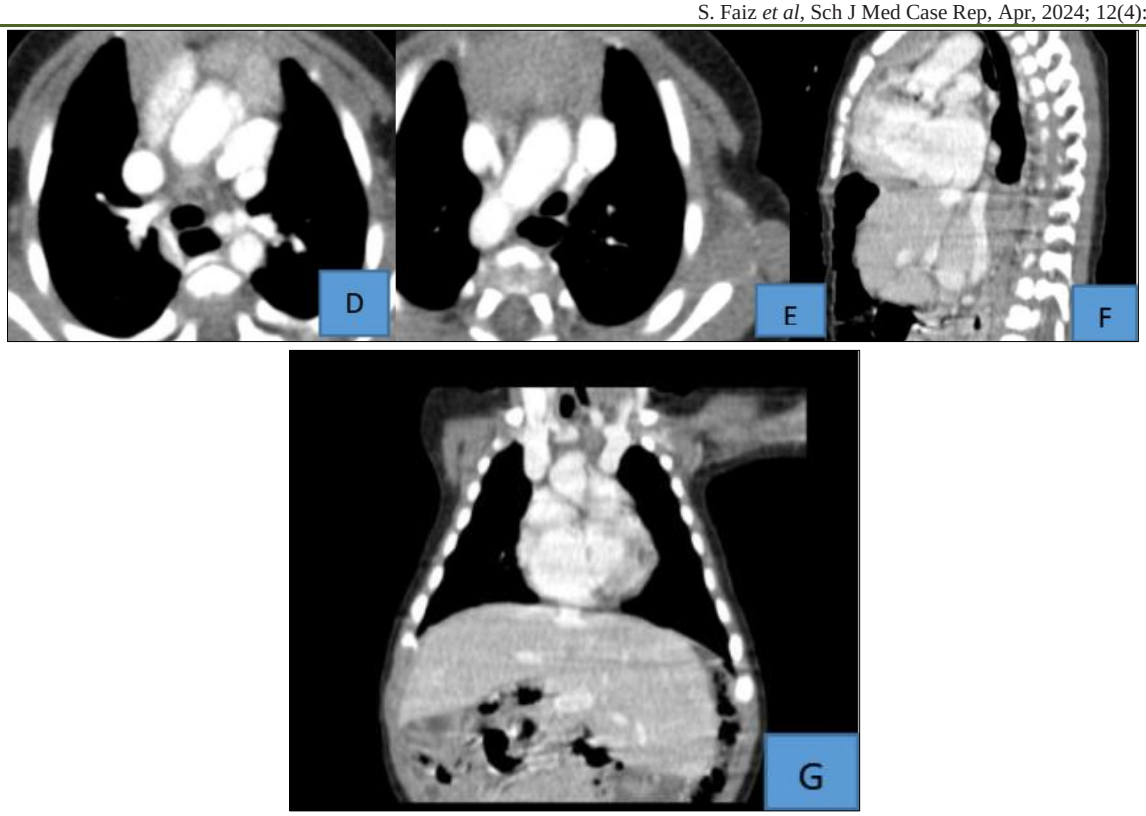
Report De Cas:

This was a 5-month-old male infant who presented with respiratory distress from birth, and in whom echo-cardiological investigation revealed complex cardiopathy associating: pulmonary artery atresia fed by a ductus arteriosus, hypoplastic pulmonary artery trunk, mitral atresia, CIA and CIV.

Thoracic angioscan was ordered showing:

- Complex malformation associating a single ventricle, a single atrium, pulmonary atresia, wide PCA and type III MAPCAs (Figures A, B and C).
- Complete abnormal venous return: supra-, intra- and infra-cardiac (Figs. D, E and F).
- Ambiguous situs with median liver and asplenia (Fig. G).





DISCUSSION

Total anomalous venous return:

The 4 pulmonary veins drain into a single collector located behind, and independent of, the left atrium. The collector then drains into different types of networks:

- Supra cardiac (type I) (40-50%):
  - ✓ Drainage from the collector to a left ascending vertical vein, then to the innominate venous trunk and superior vena cava. The vertical vein usually passes anteriorly to the hilum, or between the left pulmonary artery anteriorly and the left main bronchus posteriorly, with possible compression of the vein at this point.
  - ✓ Drainage on the right to a right ascending vein and the superior vena cava. The ascending vein may be compressed between the right pulmonary artery anteriorly, and the right bronchus or trachea posteriorly.
- Cardiac (type II) (20 to 30%): drainage to the right atrium directly or via the coronary sinus
- Infra-cardiac (type III) (10 to 30%): drainage via a vertical vessel descending through the hiatal orifice to - most often the portal system, directly or via the sinus venosus, - or the inferior vena cava or the supra-hepatic vessels. It is in these forms that there is often an obstacle to venous return: in the vertical path between the aorta and the left atrium, when crossing the diaphragm, or passing through the hepatic sinusoids, or due to closure of the sinus venosus.
- Mixed: diaphragmatic and subdiaphragmatic drainage

♣ Malformative associations: an atrial septal defect (ASD) or persistent foramen ovale is always associated, without which the malformation would be incompatible with life. Other malformative associations may be present: single ventricle, atrio-ventricular canal, tetralogy of Fallot, anomaly of the systemic venous system, left tri-atrial heart, asplenia syndrome (type III), biliary atresia, etc.

Clinical Features

cyanosis, heart failure and pulmonary hypervascularization are highly suggestive of the diagnosis. 3 types of presentation are possible:

♣ Severe pulmonary venous obstruction: neonatal forms, especially type III. Respiratory distress and severe cyanosis are prominent. Without rapid corrective surgery, the evolution is fatal

♣ Cardiac failure: on significant left-right shunt. Tachycardia, hepatomegaly, polypnea. May appear progressively in the first month as postnatal pulmonary resistance falls, explaining asymptomatic neonatal forms. No survival possible without surgical correction.

Radiologically

- Cardiac Ultrasound:

Easy-to-access examination, always performed in context. In the case of total abnormal return (where it is diagnostic), it can show:

– Absence of pulmonary veins in the left atrium.

- Presence of the retroatrial collector, stenosis of the drainage vein (acceleration of velocities).
- CIA or foramen ovale.
- Dilatation of the right atrium, ventricle and pulmonary arteries (types I, II).
- A supra- or subdiaphragmatic vein with flow away from the heart (pathognomonic of a total abnormal return, type III).
- Associated heart disease.
- Pulmonary pressures can be assessed.
- In general, it cannot be used to topograph the abnormal venous return.

- Angioscan:
  - Easy to access, especially for ventilated children, and performed without sedation, it provides a complete pulmonary and cardiovascular anatomical assessment in a single acquisition. It is particularly indicated in scimitar syndrome, where pulmonary, cardiovascular and other interrelated malformations are present.
  - It confirms abnormal venous return and the presence of the collector (which may be difficult to differentiate from the left atrium).
  - It shows the course and orientation of the draining vein.
  - It looks for compression along its path (between the left main bronchus and the pulmonary artery for type I lefts; between the aorta and the atrium, or at the level of the diaphragm for type IIIs). Stenosis may be suggested by pulmonary stasis (thickening of inter-lobular septa, ground-glass areas).
  - It provides little information on intracardiac anomalies (CIA, etc.) in the absence of gating.

EVOLUTION: 80% death within the first year in the absence of surgical treatment.

Treatment

- ♣ Prostaglandins to keep the ductus arteriosus open, surgery in all cases, as soon as diagnosis is made.
- ♣ Anastomosis of the collector with the left atrium (extracardiac forms I and III), closure of the CIA and drainage vein
- ♣ Or displacement of the inter-atrial septum so that the pulmonary vein drainage orifice is in the left atrium

CONCLUSION

Rare malformations of polymorphous clinical presentation, pulmonary venous return anomalies, whether of severe neonatal onset or in the setting of unexplained PAH, need to be understood in order to provide the best possible answers to pre-therapeutic questions, such as the anatomy of an abnormal return, its stenotic nature, and the presence of abnormal systemic vessels requiring embolization.

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