Total anomalous pulmonary venous return with mixed drainage and double connection: a rare case report not previously described

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Background
Total anomalous pulmonary venous return is a rare cyanotic congenital heart disease in which pulmonary veins connect to the systemic veins, right atrium, or coronary sinus. Given its variability, it is essential to define the pathway and drainage site.

Case summary
We describe a 3-day-old patient with total anomalous pulmonary venous return, in which the four pulmonary veins drained into a common vertical vein that had a double connection at the cardiac (coronary sinus) and infracardiac (portal vein) levels. The echocardiographic diagnosis was confirmed by computed tomography scan. At 1.5 months, she underwent surgery by anastomosis of the venous collector with the left atrium, unroof of the coronary sinus, and closure of its communication with right atrium. The vertical vein was ligated. There were no complications after 1 year of follow-up.

Discussion
This is an unusual anatomy that has not been described in the literature. In this article, we discuss the embryological origin. Furthermore, we highlight the importance of an accurate diagnosis of the venous pathway and its impact on surgical planning and patient prognosis.

Keywords
Case report • Total anomalous pulmonary venous return • TAPVR • Echocardiography

ESC Curriculum
2.2 Echocardiography • 2.1 Imaging modalities

Learning points
- Because of the variability of pulmonary venous drainage, the event may be difficult to classify or unclassifiable. We propose an accurate description based on location of the drainage, vessel connection, and pathways.
- We reflect the possibility of an echocardiographic diagnosis of the venous pathway and connections and its importance on surgical planning and patient prognosis.

Introduction
Total anomalous pulmonary venous return (TAPVR) is a rare cyanotic congenital heart disease in which pulmonary veins connect to the systemic veins, right atrium, or coronary sinus. 1

In 1957, Darling et al. proposed a classification based on the anatomical location of the drainage: supracardiac (type I), cardiac (type II), infracardiac (type III), or mixed (type IV) 1,2,3,4 (Figure 1). Because of great TAPVR variability, it is essential to define in detail the pathway, vessel connections, and drainage site. In this way, clinical evolution and surgical
implications may be predicted, allowing for an adequate management.\textsuperscript{1,3}

We present a case of TAPVR with cardiac and infracardiac drainage, with double connection diagnosed by echocardiography and confirmed by computed tomography (CT) angiogram. Venous return occurs through the confluence of the four pulmonary veins in a vertical vein that has an intracardiac connection with the coronary sinus. Infracardiac drainage results from the drainage of this vertical vein into the portal vein. This is a mixed pulmonary venous return not previously described in the literature.

**Timeline**

| Birth | Clinical picture | Cardiological examination | Angio-TC | Hospital stay | Surgery | Follow-up |
|-------|------------------|---------------------------|----------|--------------|---------|-----------|
| Asymptomatic | Physical exam: Systolic murmur (II/VI) in the left sternal border and arterial oxygen saturation ($\text{SatO}_2$) 80–85%. Chest X-ray: discrete cardiomegaly and mild venous congestion. Electrocardiogram: Normal tracing. | Echocardiographic assess | Confirmed diagnosis | Asymptomatic and haemodynamically stable | Anastomosis of retroatrial confluence to the left atrium intracardially. Unroof of coronary sinus and closure of its ostium. Ligation of the vertical vein and the patent ductus arteriosus. | Asymptomatic |
| Day 0 | Day 3 | Day 10 | Day 17 | Day 40 | Day 365 |
**Case description**

The 3-day-old patient was a newborn at term (38 weeks) weighing 2600 g, with no relevant history, who underwent a cardiological examination, due to a mesosystolic murmur (grade II/VI) in the left sternal border and desaturation of 80–85%. Chest X-ray showed discrete cardiomegaly and mild venous congestion. A normal electrocardiogram tracing was performed.

Echocardiography revealed coronary sinus dilatation and a wide atrial septal defect with a right-to-left shunt.

The echocardiography showed a retroatrial common confluence in which the right pulmonary veins and the left superior pulmonary vein converged. This confluence communicated with the coronary sinus through a 3.5-mm opening (intracardiac drainage). The confluence descended into the abdomen through a vertical vein that crossed the diaphragm and drained into the portal vein (infradiaphragmatic drainage). Before reaching the abdominal cavity, this vertical vein was joined by the left inferior pulmonary vein (Figures 2 and 3). No signs of significant pulmonary venous flow obstruction were found. A ductus arteriosus with a left-right shunt was also observed.

A thoracoabdominal CT angiogram confirmed the findings; the right and left pulmonary veins joined a common venous confluence (vertical vein) that descended behind the left atrium, draining into the portal vein. There was a small connection between this confluence and the coronary sinus. The left inferior pulmonary vein joined the confluence at the thoracoabdominal transition (Figure 4).

During her hospital stay, the patient remained haemodynamically stable. She underwent surgery at 1, 5 months, weighing 4.2 kg under extracorporeal circulation. The retroatrial confluence was anastomosed to the left atrium intracardiacally. The coronary sinus was unroofed, and the orifice of the coronary sinus, that committed to the right atrium, was closed. The vertical vein was ligated just before its passage into the abdominal cavity, as was done with the ductus arteriosus. The patient did not require inotropic support after cardiac surgery. There were no complications after 1 year of follow-up.

**Discussion**

The incidence of TAPVR is 0.008% in live births, comprising 1–1.5% of all congenital heart diseases. The distribution is quite consistent: type I (45–55%), type II (25–30%), type III (13–25%), and type IV (2–5%).

Type I consists of drainage into the systemic venous system via a left vertical vein to a left innominate vein or directly to right superior vena cava. Type II constitutes direct drainage into the heart by the coronary sinus or right atrium. Type III includes drainage by a descending vein crossing the diaphragm and joining the inferior vena cava, hepatic veins, portal system, or umbilical vein. Type IV consist of the coexistence of two or more of these drainages.

There have been reports of patients in whom pulmonary veins drain at the same anatomical level through several inflow connections. In such cases, the pulmonary veins usually drain independently to several points at the same level (e.g. infracardiac drainage where all
pulmonary veins join a common chamber that communicates independently with the portal system/venous ductus) or through a common confluence that drains into several systemic veins (e.g. vertical vein that receives all the pulmonary veins and drains infradiaphragmatic). Mixed drainage at different levels has also been described, either through independent confluences (commonly supracardiac and cardiac). More scarcely documented are the cases of supracardiac and infradiaphragmatic drainage. The anatomical characteristic of our patient, i.e. a vertical vein with simultaneous cardiac and infracardiac connection, has not been previously described in the literature.

TAPVR becomes comprehensible if embryology is considered. The precursor of pulmonary veins develops from a prominence in the left atrium. The pulmonary venous primordium extends a plexus to the pulmonary bud, which in turn connects it to the cardinal and umbilicovitelline system. Subsequently, these connections obliterate. The four major branches of the initial common pulmonary vein are embedded in the left atrium, giving rise to the normal morphology. If the interatrial septum does not develop completely or develops further to the left, some or all of the pulmonary veins will enter the right atrium and the atrial septal defect will persist. Moreover, if the pulmonary venous primordium does not join the pulmonary plexus, the venous return will occur through one of the connections with the systemic circulation, and this pathway will persist.

Apparently in our case, the common pulmonary vein failed to join the left atrium. In addition, the primitive connection to the cardinal and umbilicovitelline system persisted. This double error resulted in a common pulmonary vein that received blood from both lungs and drained into the coronary sinus and portal vein, respectively.

Currently, 2D echocardiography with colour flow Doppler performed by experienced professionals enables an accurate diagnosis. However, in cases of difficult assessment or mixed drainage, a magnetic resonance or computed tomography will provide a detailed anatomy.
The decrease in morbidity and mortality rates due to a more accurate diagnosis and improved surgical techniques has been well described.\textsuperscript{5,6} Despite the few cases reported, different risk factors have been studied. The development of pre- or post-surgical pulmonary venous obstruction (PVO) stands out as a risk factor, along with other PVO contributing factors: pulmonary vein stenosis/hypoplasia, absence of a common venous drainage, young age and low weight, and postoperative pulmonary hypertension.\textsuperscript{1,5,6}

There are no statistically significant data linking the mixed type with higher mortality. However, some studies establish infracardiac drainage as a risk factor for the development of preoperative PVO and cardiac drainage as a protective factor.\textsuperscript{1,4,5,6}

Mixed type shows a mean survival rate of 85.2\% 3 years after surgery, decreasing to 58.7\% if postoperative PVO.\textsuperscript{1}

Clinical (low weight, shock…) and preoperative morphology (narrow venous pathways or single/small venous confluence) are perceived as risk factors for survival and for development of postoperative PVO.\textsuperscript{1}

**Conclusion**

We describe an unusual case of mixed type TAPVR (cardiac and infracardiac) of a common vertical vein with a double connection to the coronary sinus and the portal vein. This work is intended to demonstrate that 2D echocardiography with colour Doppler allows accurate description of the venous pathway and drainage levels. Nevertheless, in some cases, other imaging techniques may be required for confirmation. This will allow us to make a prognostic approach and establish an individualized strategy and surgical planning.

**Lead author biography**

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**Supplementary material**

Supplementary material is available at *European Heart Journal – Case Reports* online.
Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: In accordance with COPE guidelines, the patient’s mother has signed the European Heart Journal - Case Reports patient consent form, in which she consents to the publication of the case for scientific purposes.

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Data availability
Data available on request from the authors.

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