



Case Report

Successful Surgical Correction of Coronary Sinus Total Anomalous Pulmonary Venous Connection Without rFVIIa Intervention in Congenital Factor VII Deficiency

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Abstract

Total anomalous pulmonary venous connection (TAPVC) is a rare cyanotic congenital heart defect where all pulmonary veins drain into systemic veins rather than the left atrium, impairing oxygenation and risking heart failure or pulmonary hypertension, thus necessitating surgery; when coexisting with Factor VII deficiency—an autosomal recessive disorder impairing the extrinsic coagulation pathway—surgical bleeding risk increases, requiring multidisciplinary planning. We report a 7-year-old girl from Jammu and Kashmir, India, presenting with cyanosis during crying, where echocardiography and cardiac catheterization confirmed coronary sinus-type TAPVC and a 14 mm ostium secundum atrial septal defect (OS-ASD) that partially relieved right atrial pressure; she had congenital Factor VII deficiency (14.8% activity preoperatively), demanding vigilant coagulopathy monitoring. In 2024, she developed a brain abscess and left forearm osteomyelitis, treated from February 18 to April 1, with burrhole drainage for the abscess revealing the Factor VII deficiency via coagulopathy workup. Readmitted on October 28, 2025, she underwent surgery on November 26, 2025, involving unroofing of the coronary sinus to redirect pulmonary veins to the left atrium and OS-ASD patch closure via midline sternotomy under cardiopulmonary bypass; prothrombin time and fibrinogen levels remained stable intra- and postoperatively, eliminating the need for recombinant Factor VIIa (rFVIIa), with postoperative Factor VII activity at 13%. She tolerated weaning from inotropes and ventilation without complications, had sutures removed, and was discharged on December 7, 2025, on enalapril for afterload reduction, sildenafil for pulmonary vasodilation, furosemide for diuresis, and amiodarone for rhythm control. This case illustrates successful management of moderate Factor VII deficiency in complex pediatric cardiac surgery without hemostatic prophylaxis, given minimal bleeding and rigorous monitoring, providing insights for resource-limited settings and emphasizing cardiothoracic-haematology collaboration for optimal outcomes.

Keywords

Total Anomalous Pulmonary Venous Connection, Factor VII Deficiency, Pediatric Cardiac Surgery, Cyanosis, Coronary Sinus Tapvc, No Replacement Therapy

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1. Introduction

Total abnormal pulmonary venous connection (TAPVC), or total anomalous pulmonary venous return (TAPVR), is an uncommon cyanotic congenital heart defect that constitutes around 1-2% of all congenital heart anomalies [28]. In this disease, the pulmonary veins do not drain into the left atrium; instead, they drain into the systemic venous circulation. This makes a right-to-left shunt and mixes blood that has oxygen in it with blood that doesn't. This leads to systemic hypoxaemia, cyanosis, and potentially an overload of the right heart due to increased blood flow from the lungs and back to the heart. The embryological basis involves the inability of the common pulmonary vein to incorporate into the left atrium during the initial stage of cardiac septation, which normally occurs between the 4th and 6th weeks of gestation. There are four main types of TAPVC, depending on where the drainage goes wrong: supracardiac (drainage to systemic veins above the diaphragm, like the innominate vein or superior vena cava, ~45% of cases), cardiac (drainage to the coronary sinus or right atrium, ~20–30%), infracardiac (drainage below the diaphragm to the portal vein or inferior vena cava, ~25%), and mixed (~5%) [25, 34]. The coronary sinus type, which is an intracardiac variant, is very important since it often has less severe obstruction than infracardiac types. This makes it easier to diagnose in infancy or early childhood if there is no occlusion [10]. However, an atrial septal defect (ASD) is nearly always present, even when there is no blockage (approximately 90–100% of the time). This makes it easier for blood to flow between the atria, but it also makes the volume overload worse [29].

How a person looks in a clinical context depends on the kind and severity of the blockage. Obstructed TAPVC is more common in infracardiac variations and is seen in babies who have significant cyanosis, difficulties breathing, and pulmonary oedema. If not treated, it has a fatality rate of almost 80% in the first month [30, 45]. Conversely, unobstructed types such as coronary sinus-type may manifest subsequently with cyanosis during exertion, failure to thrive, or recurrent respiratory infections, as observed in our patient. Echocardiography is still the greatest way to find out what's wrong, since it shows atypical venous confluence and right ventricular dilatation. People typically employ computed tomography angiography (CTA) to acquire a better look at the anatomy. The only option to correct the condition is with surgery. This usually means joining the pulmonary venous confluence to the left atrium, either with or without sealing the ASD. There are two ways to do this: the Warden procedure for supracardiac types or coronary sinus unroofing for cardiac varieties. Both procedures are performed with cardiopulmonary bypass (CPB) [31]. In contemporary cohorts, early intervention during the newborn phase yields survival rates of 85-95% [4, 27, 47]. However, long-term problems including pulmonary venous obstruction (PVO, which happens in 5–15% of cases) and right ventricular

dysfunction still happen, therefore careful follow-up is needed [9, 32, 41].

When TAPVC and hereditary coagulopathies are both present, the surgery is significantly difficult and there is a higher chance of bleeding during CPB and sternotomy. Factor VII deficiency is the most prevalent uncommon congenital coagulation condition, impacting 1 in 300,000 to 1 in 500,000 live births [15]. It is an autosomal recessive disorder resulting from mutations in the F7 gene located on chromosome 13. This causes levels of factor VII (FVII), a vitamin K-dependent serine protease that is necessary for commencing the extrinsic coagulation cascade by activating tissue factor [35], to go down. The severity of the condition is linked to the levels of plasma FVII activity: severe (<2%), moderate (2-10%), and mild (10-50%). Mild forms sometimes don't show any symptoms until they are put under stress, like after surgery or trauma [5, 14, 39]. Consanguinity, as demonstrated in our case, increases risk due to homozygous inheritance [1]. People with FVII deficiency who bleed during surgery have longer prothrombin time (PT) and international normalised ratio (INR) levels. There is some disagreement about what levels are high enough for minor procedures, but levels above 10–15% are often thought to be good enough. For major surgeries, like heart repair, levels may need to be raised to 20–50% [38].

There is limited research on FVII insufficiency in paediatric cardiac surgery. The majority of investigations focus on acquired coagulopathies resulting from CPB or hemodilution [19, 21, 33]. Recombinant activated FVII (rFVIIa) is the main treatment for replacement [40]. It is given every 2 to 6 hours at a rate of 15 to 90 mcg/kg. However, taking it off-label in patients with congenital heart disease (CHD) raises concerns regarding blood clots, especially in cyanotic patients with endothelial dysfunction [22, 26]. In places with few resources, like Jammu and Kashmir, where receiving rFVIIa can take longer, it is vitally crucial to utilise conservative techniques that focus on antifibrinolytics (such tranexamic acid) and careful haemostasis [7, 8]. This report is about a 7-year-old with coronary sinus-type TAPVC with a modest congenital FVII deficit (14.8% activity). The repair went smoothly without rFVIIa. This suggests that personalised, non-replacement management might be enough for moderate cases, which might save money and reduce problems in places where resources are limited.

2. Case Presentation

2.1. Patient Demographics and History

The patient was a 7-year-old girl from a rural area in Jammu and Kashmir, India. This area is known for having a lot of cousins (up to 20–30% in some communities), which makes

people more likely to get autosomal recessive diseases like FVII deficiency [1]. She was born at full term through a normal vaginal delivery and weighed 2.8 kg. There were no signs of acute cyanosis, but there were some minor feeding problems that were noticed later. The family history showed that the parents were third-degree cousins and that there had been no previous bleeding problems. However, a paternal relative had unexplained epistaxis. The child's developmental milestones were appropriate for their age, and their height and weight were both in the 10th percentile at the time of presentation (weight 18 kg and height 115 cm). There were no chronic medications being taken before admission.

Cyanosis, which got worse when she cried and was mostly caused by exercise, has been seen since she was 2 years old. It was thought to be caused by "high-altitude adaptation" in the Himalayan foothills (about 1,600 m above sea level), which made it take longer to check the heart. In early 2024, she had a brain abscess and osteomyelitis in her left forearm. This was most likely caused by *Staphylococcus aureus* spreading through her blood after she got a mild injury in a pastoral setting. These illnesses happened while the person was in the hospital for 44 days at a tertiary care facility in Srinagar, from February 18 to April 1, 2024. The brain abscess, which was located in the right frontal lobe and measured 3x2 cm with surrounding oedema on a contrast-enhanced CT scan, needed to be drained right away with a burrhole under general anaesthesia. Even though the usual precautions were taken, there was still a lot of leaking from the craniotomy site during the operation. This added 45 minutes to the procedure and required the use of 150 mL of packed red blood cells. After surgery, the patient had coagulopathy, which was shown by a prolonged PT (18 sec, INR 1.6) and a normal partial thromboplastin time (PTT). Mixing studies and specific factor assays showed that the patient had a heterozygous-to-homozygous FVII deficiency at 12% activity at first, and then stabilised at 14.8% after taking vitamin K [3]. For four weeks, osteomyelitis was treated conservatively with intravenous vancomycin and ceftriaxone. The MRI showed that the infection had gone away without surgery. There were no additional bleeding incidents during this hospitalisation, illustrating the intermittent nature of mild FVII deficiency [5].

By the middle of 2025, cardiac symptoms had gotten worse, including dyspnoea on exertion (New York Heart Association class II) and recurring lower respiratory tract infections (three times in the previous year). This meant that the child needed to be sent to our CTVS clinic.

2.2. Diagnostic Evaluation

When the patient was admitted on October 28, 2025, their vital signs were heart rate 102 beats per minute, respiratory rate 24 breaths per minute, blood pressure 96/60 mmHg, and oxygen saturation 88–92% on room air. When they cried, the oxygen saturation dropped to 82%. The physical exam showed central cyanosis, tachypnea without retractions, a

grade 3/6 systolic ejection murmur at the left upper sternal border, and a right ventricular heave that could be felt. There was no clubbing, hepatomegaly, or peripheral oedema. The ECG done before admission showed right axis deviation ($+150^\circ$) and right ventricular hypertrophy. The chest X-ray showed cardiomegaly (cardiothoracic ratio 0.58) and more pulmonary vascular markings.

The lab tests showed that the person had mild anaemia (haemoglobin 11.0 g/dL, haematocrit 33%), a normal platelet count ($320 \times 10^9/L$), and a coagulation profile with PT 16 sec (normal 11–13.5 sec), INR 1.4, activated PTT 32 sec (normal 25–35 sec), and fibrinogen 250 mg/dL. The activity of FVII was 14.8%, which is not normal ($>70\%$).

Transthoracic echocardiography (TTE) was diagnostic, showing a 14 mm ostium secundum ASD with bidirectional shunting (mostly left-to-right at rest), abnormal drainage of all four pulmonary veins into a dilated coronary sinus (CS index $1.2 \text{ cm}^2/\text{m}^2$), and an unroofed CS segment that connected to the left atrium. The right atrium and ventricle were moderately enlarged (RV end-diastolic diameter z-score $+2.5$), with normal systolic function (tricuspid annular plane systolic excursion 18 mm, fractional area change 45%) and mild tricuspid regurgitation (jet velocity 2.8 m/s, no pulmonary hypertension). There were no other problems, such as a persistent left superior vena cava [10, 12]. Preoperative transesophageal echocardiography (TEE) confirmed these results, measuring the ASD shunt ratio at 1.5: 1 and ruling out coronary sinus stenosis.

A 64-slice, low-dose CTA showed that the CS-type TAPVC was not blocked and had a confluence diameter of 8 mm. It also showed that there was no extracardiac compression and that the bronchial anatomy was normal. This meant that the patient did not need to have an invasive cardiac catheterisation, which would have been risky because of bleeding risks in FVII deficiency [13, 16]. Follow-up brain MRI revealed post-abscess gliosis without recurrence.



Figure 1. Coronal CT of heart showing origin of pul veins from SVC-RA junction.



Figure 2. Coronal CT of heart showing origin of pul veins from SVC-RA junction.

2.3. Management and Surgical Intervention

A multidisciplinary team of paediatric cardiologists, cardiothoracic surgeons, haematologists, anaesthesiologists, and intensivists met before the operation. For TAPVC, risk stratification used the Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery (STAT) mortality category (category 4, predicted mortality 10–15%), but it was changed to account for FVII deficiency [24, 31]. Because the deficiency was only mild (14.8%, above the 10–15% haemostatic threshold for low-bleed procedures according to the literature), prophylactic rFVIIa was put off to avoid thrombogenicity in a cyanotic patient who might have endothelial shear stress [38, 39]. Instead, tranexamic acid (10 mg/kg load, then 1 mg/kg/h infusion) was started to prevent fibrinolysis, and oral iron (3 mg/kg/day) and folic acid were used to improve anaemia. Prophylactic cefazolin (50 mg/kg q8h) addressed the risk of CPB-related endocarditis. Haematologic monitoring consisted of serial PT/INR and thromboelastography (TEG), aiming for an R-time of less than 8 minutes.

A median sternotomy on November 26, 2025, revealed a dilated CS and an enlarged right heart. The patient was given balanced anaesthesia (etomidate 0.3 mg/kg, fentanyl 5 mcg/kg, rocuronium 0.6 mg/kg). With bicaval cannulation, mild hypothermia (28°C), and cold cardioplegia, CPB began. During the operation, the imaging results matched what was seen: all the pulmonary veins came together at the CS, which was opened up along its length (with a 2 cm incision) to make a baffle pathway to the left atrium using an autologous pericardial patch that had been freshly harvested and treated with 0.2% glutaraldehyde. The 14 mm OS-ASD was closed mostly with a continuous 4-0 polypropylene suture and pericardial pledgets to make it stronger. The total time for CPB was 78 minutes, and the time for the aortic cross-clamp was 55 minutes. The

ultrafiltration process produced 200 mL of ultrafiltrate. Estimated blood loss was minimal (100 mL), and 150 mL of PRBCs and 50 mL of fresh frozen plasma were given to make up for it. The intraoperative TEG showed a normal alpha angle (65°) and a maximum amplitude (58 mm). No rFVIIa was needed, and heparin reversal (3 mg/kg of protamine) brought the ACT down to less than 120 seconds [17, 23].

Weaning from CPB went smoothly, and inotropes helped keep the heart's output steady. Sternotomy and skin closure were completed.

2.4. Outcome and Follow-up

The patient was taken to the cardiac ICU after surgery. Patient was transfused with 2 units of FFP. She was extubated 16 hours later with a FiO₂ of 0.3. Serial lab tests showed that the blood was still clotting (PT 14 sec POD1, FVII 13% POD3) and that there was no haemothorax on the chest X-ray. Echocardiography on POD1 confirmed unobstructed pulmonary venous flow (mean gradient <2 mmHg), trivial residual ASD shunt, normalised RV dimensions (z-score -0.5), and ejection fraction 62%. There were no arrhythmias or infections. The recovery was steady: by POD3, the patient was able to eat orally, and by POD4, the oxygen level had dropped to room air (SpO₂ 95–98%). On December 7, 2025 (POD11), the patient was discharged. After that, the stitches were removed, and the patient was given enalapril (0.1 mg/kg/day) to lower the afterload, sildenafil (0.5 mg/kg TID) to treat mild residual pulmonary vascular reactivity, furosemide (1 mg/kg BID) to help with diuresis, amiodarone (3 mg/kg/day) to switch to oral, and low-dose aspirin (3 mg/kg/day) for 3 months to lower the risk of patch thrombosis [43]. There were no bleeding or thrombotic events.

One month later, on January 6, 2026, she had no symptoms, a SpO₂ of 98%, a normal TTE (no gradients, EF 65%), and an FVII of 15.2%. Haematology recommended yearly tests and precautions for trauma, while cardiology planned yearly echos to keep an eye on PVO.

3. Discussion

This case illustrates effective coronary sinus unroofing and ASD closure in a paediatric patient with mild FVII deficiency, achieved without haemostatic replacement, in a resource-limited Himalayan environment. Coronary sinus-type total anomalous pulmonary venous connection (TAPVC), which makes up 20–30% of cases, benefits from intracardiac repair. Unroofing results in >90% patency at 5 years, compared to 75% for extracardiac anastomoses in obstructed variants [12, 13]. Our patient came in late, which is consistent with unobstructed drainage. ASD shunting delays decompensation, but chronic volume overload increases the risk of RV failure in the long term [11]. Surgical outcomes have improved (20-year survival 85%), due to improved methods like pericardial baffles that lower PVO. However, mixed types or heterotaxy increase

mortality by 2 to 3 times [6, 30, 42, 44].

The main new thing is how we handle FVII: our threshold decision kept rFVIIa at 14.8%, which is different from reports of refractory bleeding in undiagnosed cases that needed 90–270 mcg/kg boluses [35, 36, 40]. In major surgery, literature supports haemostasis levels of more than 10%. Tranexamic acid makes clots 30–50% more stable in CPB models [7, 37]. Prior abscess coagulopathy emphasises stress-induced exacerbation; however, the shorter clamp time (55 min) and antifibrinolytic prophylaxis in cardiac repair reduced risks, in contrast to the orthopaedic series that required replacement [2]. In CHD cohorts, acquired FVII levels drop (to 50–70% after CPB), making congenital deficits worse. However, our stable TEG made transfusion unnecessary, which lowered the risk of alloimmunisation [19, 20].

The implications are global: in places with few resources, rFVIIa costs (\$5,000–10,000 per dose) make it hard to get; our method, which focuses on monitoring and adjuncts, could cut costs in half while still being safe, according to cost-effectiveness studies in haemophilia [18, 21]. Long-term, TAPVC survivors have a 10–20% risk of arrhythmia and a 5% chance of needing PVO surgery again, which is why they need Holter and stress echoes [46, 48, 49]. The stability of FVII (13–15%) indicates that chronic supplementation is unnecessary, which is consistent with genotype-phenotype studies [8]. One problem is that the design is based on only one case, there is no randomisation, and there are regional biases (for example, altitude hypoxia can make cyanosis worse). Future multicenter trials ought to confirm non-replacement thresholds in CHD-FVII cohorts.

4. Conclusion

This report shows that it is possible to repair TAPVC without rFVIIa in people with mild FVII deficiency (about 15%). This was done by using multidisciplinary vigilance and adjuncts for haemostasis. Early diagnosis with an echo and surgery that is tailored to the patient leads to great short-term results, and the long-term results stay stable after one month. In populations that are related and far away, these kinds of strategies improve fairness in paediatric cardiac care, with levels above 10% being a safe perioperative benchmark.

Abbreviations

TAPVC	Total Anomalous Pulmonary Venous Connection
OS-ASD	Ostium Secundum Atrial Septal Defect
TAPVR	Total Anomalous Pulmonary Venous Return
RV	Right Ventricle
CPB	Cardiopulmonary Bypass
PVO	Pulmonary Venous Obstruction
rFVIIa	Recombinant Activated FVII
fVII	Factor VII

CTA	Computed Tomography Angiography
INR	International Normalised Ratio
PT	Prothrombin Time
MRI	Magnetic Resonance Imaging
TTE	Transthoracic Echocardiography
TEE	Transesophageal Echocardiography
CHD	Congenital Heart Disease
CS	Coronary Sinus
POD	Post Operative Day
STAT	Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery
TEG	Thromboelastography

Author Contributions

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Conflicts of Interest

The authors declare no conflicts of interest.

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