

# Low Oxygen Saturation Following Total Correction in a Patient with Tetralogy of Fallot and Persistent Left Superior Caval Vein – How Did We Diagnose and Manage?

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## SOUHRN

Souvislost mezi Fallotovou tetralogií (tetralogy of Fallot, TOF) a jinými intrakardiálními patologiemi, jako jsou defekt septa síní (atrial septal defect, ASD), defekt síňokomorového kanálu nebo perzistentní levostranná horní dutá žíla (persistent left superior vena cava, PLSVC) i absence plicní chlopně, je dobře známa. Během chirurgického výkonu vyžadují přidružená onemocnění obzvláštní pozornost.

V tomto článku popisujeme léčbu čtyřměsíční dívky s diagnózou TOF a PLSVC, u níž však byl přehlédnut nezastřešený koronární sinus. Nezastřešený koronární sinus se vyskytuje současně s TOF velmi vzácně.

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## ABSTRACT

Association of tetralogy of Fallot (TOF) with the other intracardiac pathologies such as atrial septal defect (ASD), atrioventricular canal defect or persistent left superior vena cava (PLSVC), absent pulmonary valve are well known pathologies. The associated pathologies require specific attention during surgical treatment.

In this manuscript, we present management of a four-month-old girl who was diagnosed with TOF and PLSVC but the diagnosis of unroofed coronary sinus was missed in her. Association of unroofed coronary sinus with TOF is a very rare variant of TOF pathology.

## Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease.<sup>1</sup> It is accounted 5–7% among all congenital heart defects. Many congenital cardiac disorders are accompanied by persistent left superior vena cava (PLSVC) combined with large coronary sinus.<sup>2</sup> However, unroofed coronary sinus (UCS) is quite a rare variant among the atrial septal defects. It is much rarer that this very rare pathology accompanies other cardiac pathologies. PLSVC cases accompanying TOF are known to exist, although rarely seen.<sup>3–5</sup>

A 4-month-old girl was admitted to our institution with the diagnosis of TOF and PLSVC. Unroofed coronary sinus was detected during intraoperative echocardiography which was performed due to persisting low oxygen saturation despite total correction. In this paper, we aimed to present the management of the patient.

## Case report

A 4-month-old girl with the diagnosis TOF was referred to our clinic for surgical treatment. She weighed 5 600 g

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and was 56 cm tall. Her room air saturation was 84%. Echocardiography (ECHO) showed findings compatible with TOF accompanied by PLSVC and a large coronary sinus. A large VSD was present in perimembranous area extending to the outlet, and also infundibular, annular and valvular pulmonary stenoses were present in echocardiography. The aortic arch was located on the right side. In echocardiographic evaluation the McGoon ratio was above 2, there was no additional VSDs and coronary arteries were normal and total correction operation was decided for the patient after informing the patient's family about the operation in details and their consent.

With general anesthesia and standard median sternotomy measures, aortic, and trival cannulation was performed. Following cross clamp VSD was explored through right atriotomy, and closed with gluteraldehyde treated pericardial patch with interrupted pledgeted sutures. Arteriotomy was performed on the main pulmonary artery. Pulmonary valve was bicuspid and annular hypoplasia was present. Incision was extended toward the right ventricular infundibulum while protecting the valves. Hypertrophic muscle bands on the right ventricular outflow tract were divided. The right ventricular outflow tract was reconstructed with transannular monocusp autologous pericardial patch. Cardiopulmonary bypass was completed uneventfully but oxygen saturation was monitored to be between 45–50% despite inspiratory 100% oxygen inhalation. The right ventricular / left ventricular systolic pressure ratio was found 0.42. Both lungs were functioning well and end-expiratory pressure and end-tidal carbon dioxide levels were within normal limits. Transcardiac echocardiography was performed and it showed no residual ventricular septal defect, no obstruction at the right ventricular out flow tract. The echocardiography showed surprisingly that coronary sinus was large and there was bidirectional communication between left atrium and the coronary sinus (Fig. 1). Following cardioplegic arrest, right atriotomy was performed again. Foramen ovale was expanded. The atrial septal defect was closed with autologous fresh pericardial patch so that the coronary sinus drained into the right atrium. Cardiopulmonary bypass was successfully completed with the help of 0.5 µg/kg/min milrinone and 0.05 µg/kg/min adrenaline inotropic supports. The oxygen saturation was

monitored as 97–99%. The operation was duly completed. The patient was extubated at the 14th postoperative hour. Oxygen saturation was also monitored as 97–99% in the following days. After two days of intensive care and 6 days of follow-up in the ward, the patient was discharged without any problems. The case has been followed up uneventfully for more than 8 months.

## Discussion

Tetralogy of Fallot is the most common cyanotic congenital heart disease in children.<sup>1</sup> The presence of PLSVC with the enlargement of the coronary sinus is frequently encountered in isolation or associated with other pathologies. However, UCS type ASDs are quite rare. If they are not evaluated in details, they can be easily missed out in pediatric cardiology and cardiovascular surgery practice. Payne et al. reported that they detected UCS during the aortic valve replacement operation of a patient.<sup>6</sup> Niyogi et al. found UCS in a patient who underwent double outlet right ventricle treatment during the examination they carried out due to post-operative hypoxia.<sup>7</sup> Yarrabolu et al. reported that they found UCS in a 6-month-old boy who developed cyanosis after surgical closure of VSDs.<sup>8</sup> Mallula et al. reported that they successfully completed the PLSVC and coronary sinus ASD in a 2-month-old case with TOF.<sup>3</sup> Ramman et al. found UCS in a patient with operated TOF and PLSVC who developed post-operative hypoxia.<sup>4</sup>

Persistent left superior vena cava is frequently diagnosed in centers concentrated on congenital heart diseases. However, coronary sinus pathologies can be missed out and found later either as a result of complications or through post-operative desaturation. Alertness is necessary for coronary sinus defects particularly in the presence of PLSVC. In addition, in case cyanosis continues after the corrective procedure in patients with cyanotic congenital heart diseases like TOF, rare coronary sinus anomalies should be considered if there are no other pathologies to explain the postoperative cyanosis.

The diagnosis of uncommon pathologies associated with TOF may change surgical strategy. If the pathology causes hemodynamical instability, it is better to handle the pathology during intracardiac repair of TOF. If there is PLSVC cannulation must differ from usual TOF patients. In this patient we also cannulated knowing that patient have associated PLSVC. But here since we were not informed about the inter-atrial communication through unroofed coronary sinus we failed to inspect the intracardiac anatomy in details at first. When the perioperative echocardiography revealed unroofed coronary sinus due to persisting low oxygen saturation, we went on cardiopulmonary bypass and managed the pathology.

If oxygen saturation is found to be low and if it is difficult to wean off cardiopulmonary bypass after total correction operations of cyanotic congenital heart diseases, certain issues should be checked routinely. We must be sure that there is no problem with ventilation blood gases and end-expiratory carbondioxide and pressures must be within normal limits. Also intraoperative TEE must be performed routinely to see any misdiagnosed intracardiac pathologies in these cases.

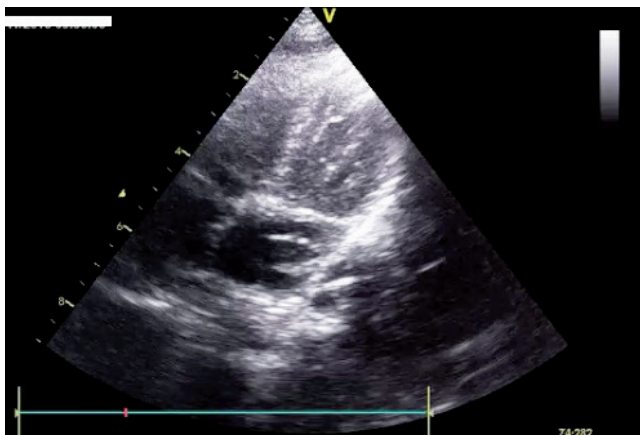


Fig. 1 – Echocardiographic view of unroofed coronary sinus.

As a general medical knowledge, it is known that unroofed coronary sinus may cause cyanosis. In cases of TOF, where cyanosis is a natural result of pathology and clinical presentation, it is understandable that other possible causes of this cyanosis may be overlooked before the operation. As in our particular case, the preoperative desaturation was relied on the diagnosis of TOF. However, it persisted after the closure of interventricular and interatrial communications when we first weaned off cardiopulmonary bypass. The degree of desaturation is dependent to the right-to-left shunt, which, in turn, links to the amount of systemic venous blood carried by the PLSVC and the proportion of systemic venous blood that crosses the atrial septum and reaches the systemic circulation. In cases of TOF since RV compliance is not decreased if there is any interatrial communication, usually the blood shunt is mainly from right to left after the operation. In some cases the right superior vena cava is rudimentary the PLSVC is more prominent, in these cases also the cyanosis could be more than expected.

In conclusion, with our particular case we wanted to emphasize a few important issues during cardiac surgery; 1: pediatric TEE must be present for all age and weight patients in pediatric cardiac surgery department, 2: UCS should be kept in mind in cases where cyanosis continues after total correction operation, 3: association of unroofed coronary sinus with TOF is extremely rare.

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