Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

A 4-month-old male with Raghib syndrome: a rare case report from Syria

Mouhammed Sleiay, MD[a,*], Bilal Sleiay, MD[a], Obeda Batrash, MD[a], Hadi Alabdullah, MD[a], Hasan Alsmoudi, MD[a], Douha AlBaroudi, MD[a], Ahmad Almohamed, MD[a], Mohammad Ali Abshi, MD[a], Saleh Takkem, MD[b]

Introduction and importance: Raghib syndrome is a rare congenital complication consisting of the termination of the left superior vena cava (LSVC) in the left atrium, an unroofed coronary sinus, and an atrial septal defect most often found in the posterior-inferior angle of the atrial septum. Both a right-to-left and a left-to-right intracardiac shunt exist. In most circumstances, they do not show any symptoms.

Presentation of case: The patient presented with a persistent left superior vena cava draining into the left atrium, an unroofed coronary sinus, and a secondary atrial septal defect (ASD). Transthoracic echocardiography was used to diagnose the condition, and surgery was applied as the primary treatment.

Clinical discussion: It was formerly believed that this complex was exclusive to Raghib syndrome; however, cases have been found in which the interatrial connection is the aperture of the unroofed coronary sinus in patients with a normal atrial septum.

Conclusion: Extracardiac treatment for this illness reduces the load on the left atrial suture and may stop further arrhythmias. There is no possibility of pulmonary vein flow restriction when there are no atrial tunnels.

Keywords: persistence of the left superior vena cava, Raghib Syndrome, tetralogy of Fallot, unroofed coronary sinus syndrome

Introduction

Raghib syndrome is a rare developmental complex that coexists with coronary sinus (CS), which is not detectable by echocardiography, atrial septal defect, and persistence of the left superior vena cava (PLSVC) canal, which is detectable by echocardiography. The PLSVC always drained into the position between the base of the left atrial appendage and the ostium of the left superior pulmonary vein in the short-axis view of the aortic root, while the proximal segment of the PLSVC was depicted going down at the left side of the aorta in the suprasternal views[1]. PLSVC anomalies comprise 2–5% of congenital heart diseases; they are usually combined with other cardiac abnormalities, but they can exist in an isolated form. According to Studer and colleagues, 3% of their atrioventricular septal defect (AVSD) patients have PLSVC with completely unroofed coronary sinus syndrome[2]. Due to the LSVC's connection to the left atrium, Raghib syndrome causes a significant left-to-right shunt through the coronary sinus defect and arterial desaturation, with a risk of paradoxical embolism. Cross-sectional imaging enables a finer characterization of the surgery's anatomy. Transthoracic echocardiography offers a sufficient diagnosis of Raghib syndrome. Desaturation and a left-right shunt are caused clinically, and they are typically accompanied by other cardiac problems, including partial or full atrioventricular channel, tetralogy of Fallot, interventricular communication, heterotaxia syndrome, and double outlet right ventricles, among others[3].

Heterotaxy syndrome and unroofed coronary sinus syndrome (UCSS) are two distinct clinical conditions in which a persistent left superior vena cava (LSVC) can drain into the left atrium (LA) in the absence of coronary sinus[4].

In these cases, biventricular repair can be done by intracardiac rerouting or by disconnection—reconnection to the right atrium (RA) or to the right superior vena cava (RSVC) using extracardiac procedures. From 1998 to 2016, biventricular repair surgeries were performed on 10 patients with LSVCs draining into the

HIGHLIGHTS

- A persistent left supraorbital vena cava and an unroofed coronary sinus characterize the uncommon heart abnormality known as Raghib syndrome.
- In 0.3–0.5% of healthy individuals and 2.1–4.3% of patients with congenital heart disease, left superior vena cava persistence is reported.
- Treating this condition extracardiacally reduces the strain in the left atrial suture and may prevent further arrhythmias.

[a]Faculty of Medicine, Hama University and [b]Department of Cardiology, Alwatani Hospital, Hama, Syria

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article

*Corresponding author. Address: Faculty of Medicine, Hama University, Hama, Syria. Tel : +963 932 624 542. E-mail: abdmouh1234mouhmouh@gmail.com (M. Sleiay).

Annals of Medicine & Surgery (2024) 86:1687–1690

Received 2 November 2023; Accepted 27 December 2023

Published online 8 January 2024

http://dx.doi.org/10.1097/MS9.0000000000001693

LA[5]. Herein we report a rare case of a 4-month-old male patient with recurrent pulmonary infections, only to find later that he has Raghib syndrome.

Case presntation

Manuscript revised final

A 4-month-old male patient was referred to the cardiology department by the paediatrician for recurrent pulmonary infection more than three times during the last three months because there was a systolic murmur in the pulmonic area. There was no obvious cyanosis. The infant had a family history of an ostium secundum defect treated surgically in one of his sisters. The lab results were as follows: White blood cells (WBCs) were 10 000, the erythrocyte sedimentation rate (ESR) was 18, blood glucose was 66 mg/dl, total calcium was 8.5 mg/dl, creatinine was 0.6 mg/dl urea was 10 µg/dl, haemoglobin was 11 g/dl, and C-reactive protein was 20 mg/dl. Sodium and potassium electrolytes were within the normal range. The electrocardiogram revealed a right-axis deviation with a right bundle branch block. The existence of pulmonary vascular prints was found via the chest X-ray. The transthoracic echocardiogram demonstrated a coronary sinus-type ASD (with additional secondum ASDs) (Fig. 1) with a dilation of both the right atrium and . right ventricle (Figs. 2, 3) and a persistent left superior vena cava (Fig. 4).

The patient was referred to the Pediatric Cardiac Surgery Center, as the treatment is surgical by closing the atrial septal defects. Both atrial septal defects—secondary ASD and unroofing coronary sinus—were surgically patched together to be fixed. As it has no negative effects on survival rates, we continued on without making any modifications to the LSVC. The child recovered after surgery without incident. The echocardiogram revealed no systemic desaturation and normal mitral valve

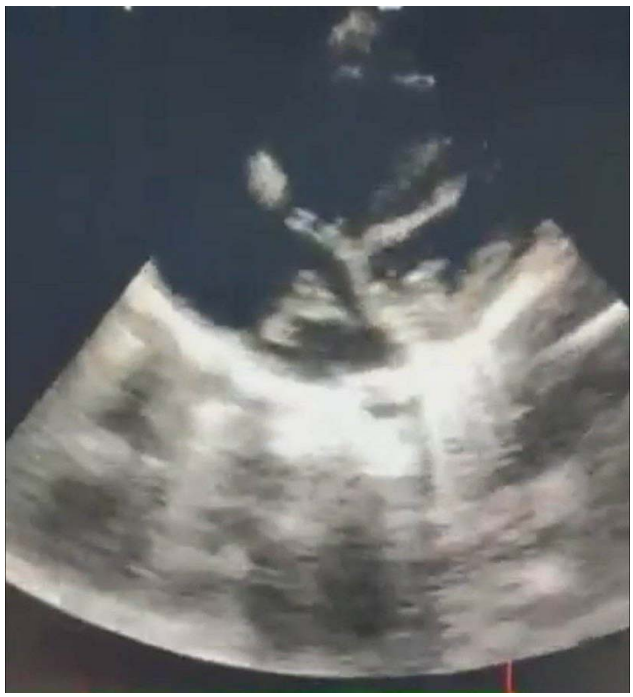

Figure 2. Apical quadrilateral section showing dilatation of the ventricle and right atrium.

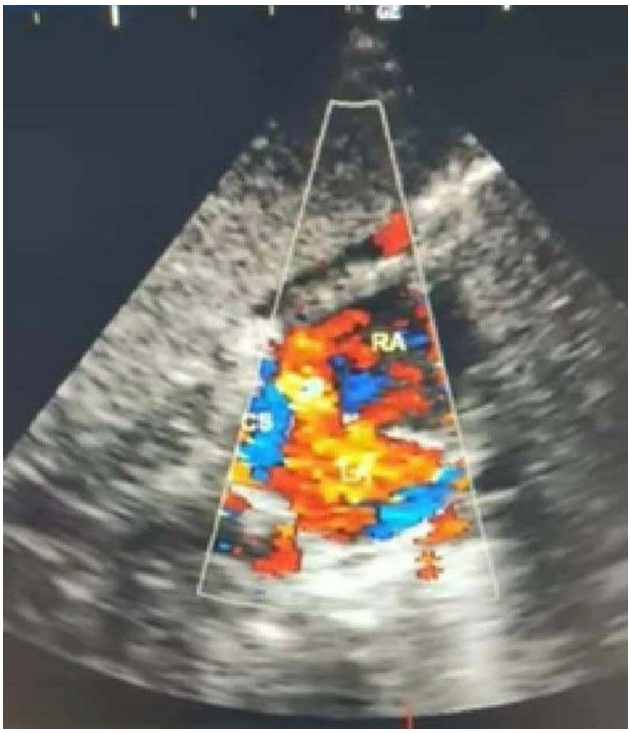

Figure 1. Substernal section showing the opening between the two atria in the coronal sinus pattern/arrow/. CS, coronal sinus; LA, left atrium; RA, right atrium.

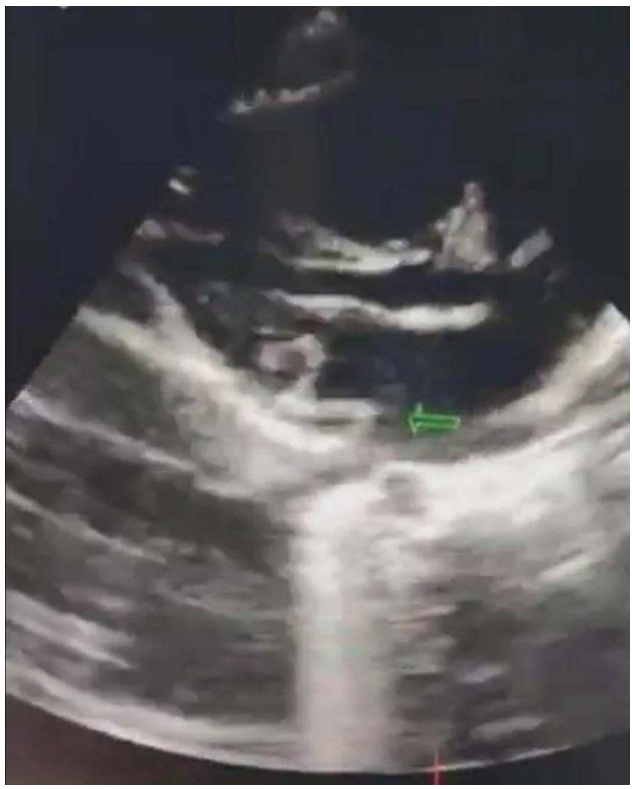

Figure 3. Parasternal cut plax shows enlargement of the coronal sinus/arrow/.

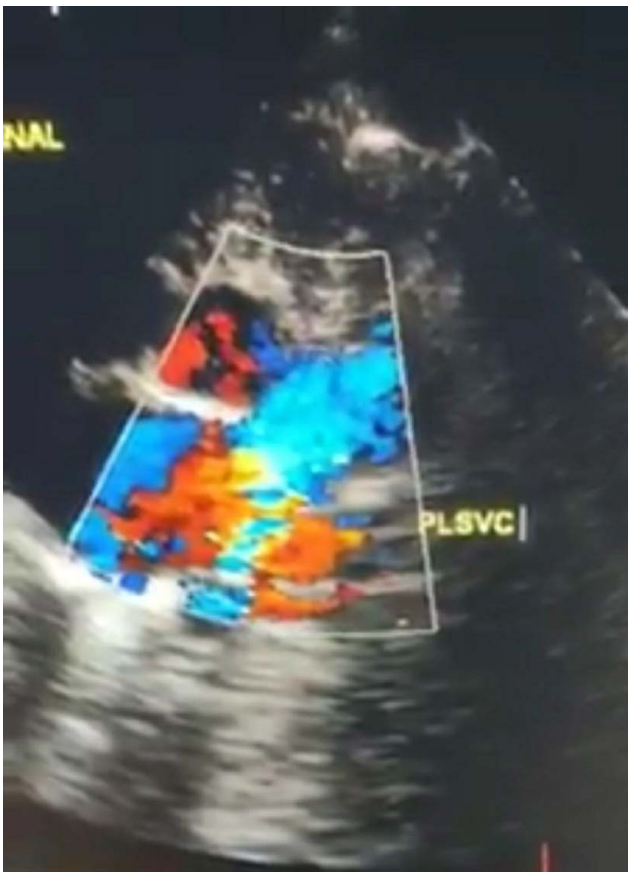

Figure 4. Suprasternal section showing the left upper accessory cavity. PLSVC, persistence of the left superior vena cava.

function. The patient's general health was satisfactory after 3 years of follow-up, and no murmurs or heart complaints were noticed.

Discussion

Raghib syndrome is a rare cardiac defect with an unroofed CS and a PLSVC[6]. Since it was originally reported in 1738, atresia of the coronary sinus ostium without this inappropriate connection is infrequent. PLSVC persistence is observed in 0.3–0.5% of normal persons and in up to 2.1–4.3% of congenital cardiac disease patients. Raghib and colleagues first described Raghib syndrome as a developmental complex that includes the left superior vena cava terminating in the left atrium in 1965[7]. In the overall healthy population, the incidence of LSVC ranges from 0.2 to 3% (2). 90% of the time, the LSVC empties its blood into the RA through the CS and has no clinical effects[8]. In 30% of cases, the innominate vein that connects them is present in this circumstance. This type of venous anomaly is more frequently associated with other cardiovascular anomalies. such as pulmonary vein anomalies (which occur in 10% of cases), pulmonary atresia (which occurs in 6% of cases), tricuspid atresia (which occurs in 4–5% of cases), hypoplasia of the left ventricle (which occurs in 2.5% of cases), and fallot tetralogy (5% of cases)[9]. Raghib and colleagues first described Raghib syndrome as a developmental complex in 1965. Raghib syndrome is a rare cardiac anomaly characterized by the left superior vena cava terminating in the left atrium, the absence of the coronary sinus, and a common atrial septal defect at the posterior-inferior angle of the atrial septum[10]. This complex was formerly thought to be exclusive to Raghib syndrome; however, examples with a normal atrial septum have been discovered in which the aperture of the unroofed coronary sinus serves as the interatrial connection[11]. In our patient, An electrocardiogram (ECG) revealed a right-axis deviation with signs of a right bundle branch block. The existence of pulmonary vascular prints was found via the chest X-ray. The patient underwent an ultrasound imaging investigation, which revealed the presence of a four-chamber echocardiography with right ventricular and right atrial dilatation as well as coronary sinus dilatation. We suggested possible differential diagnoses, such as pulmonary hypertension, congenital heart defects, and cardiomyopathy. However, the presence of a left superior vena cava anomaly as well as a coronary sinus atrial septal defect led us to the conclusion that our patient had Raghib syndrome. Numerous intracardiac and extracardiac surgical techniques have been described[12,13]. A thorough knowledge of all of these techniques is required for the appropriate selection to be made because it depends on important characteristics such as the patient's anatomy, age, and concomitant cardiopathies and abnormalities. Reroofing of the coronary sinus is focused on the creation of a new coronary sinus using the pericardium, synthetic materials, or even autologous left atrial tissue. Then, a new atrial septation must be created with a new patch[14,15]. Of the extracardiac repair methods, direct anastomosis is preferred because it maintains growth potential and has lower thrombotic complications than synthetic grafts; however, the LSVC has to be long enough to avoid any tension on the anastomosis. Techniques include anastomosing to the right atrial area (RAA), tunnelling through the transverse sinus to the RSVC, and creating a bidirectional left superior cavopulmonary anastomosis with the pulmonary artery[12]. The use of a conduit is technically easier because it does not require cross-clamping and avoids a thorough dissection of the LSVC, removing the risk of phrenic nerve injury[3]. In our situation, both atrial septal defects—secondary ASD and unroofing coronary sinus—are surgically patched together to be fixed. As it has no negative effects on survival rates, we continue on without making any modifications to the LSVC. After 3 years of follow-up, the patient's general health condition was good, and no murmurs or heart symptoms were observed.

Conclusion

Raghib syndrome is a very uncommon congenital cardiac abnormality that is defined by the lack of the coronary sinus and prolonged LSVC draining into the left atrium. There are several methods to reroute the LSVC flow to the right atrium, despite the fact that few examples have been documented in the literature. These procedures include repositioning the interatrial septum, extracardiac mobilization of the anomalous vein, and surgery to establish intracardiac flow using atrial patches to form a tunnel (along the roof of the left atrium or the theoretical course of the coronary sinus). The extracardiac method of treating this illness lowers left atrial suture tension and may stop further arrhythmias. The potential blockage of pulmonary vein flow is avoided when atrial tunnels are absent. Practioners should take Raghib syndrome into consideration for every child who has recurrent

respiratory infections, especially if he has murmurs or cadiac symptoms.

Methods

The work has been reported in line with the SCARE criteria[16].

Ethical approval

It is not applicable because all data belong to the authors of this article.

Consent

Written informed consent was obtained from the patient's parents/legal guardian for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Sources of funding

None.

Author contribution

M.S. performed the histological examination and was a major contributor in writing the manuscript. B.S. was a major contributor in writing the manuscript. O.B. was a major contributor in writing the manuscript and prepared all figures. H.A., Hasan A., D.B., M.A., A.A., S.T. wrote a part of the manuscript. All authors read and approved the final manuscript.

Conflicts of interest disclosure

None.

Research registration unique identifying number (UIN)

None.

Guarantor

Not applicable . All data belong to the authors.
The guarantor author is Mouhammed Sleiay.

Data availability statement

Not applicable because all data belong to the authors of this article.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Acknowledgements

The authors appreciate the efforts and the help of SMSR lab. and its role in binging our team together.

References

[1] Xie MX, Yang YL, Cheng TO, *et al*. Coronary sinus septal defect (unroofed coronary sinus): echocardiographic diagnosis and surgical treatment. Int J Cardiol 2013;168:1258–63.
[2] Baek WK, Kim YS, Yoon YH, *et al*. Cor triatriatum with Raghib complex in partial atrioventricular septal defect and common atrium: a rare combination. Gen Thorac Cardiovasc Surg 2020;68:641–3.
[3] Hussein N, Kasdi R, Coles JG, *et al*. Use of 3-dimensionally printed heart models in the planning and simulation of surgery in patients with Raghib syndrome (coronary sinus defect with left superior vena cava). JTCVS Tech 2020;2:135–8.
[4] Aguilar JM, Rodríguez-Serrano F, Ferreiro-Marzal A, *et al*. Left superior vena cava draining into the left atrium: Clinical entities, diagnosis and surgical treatment. Arch Cardiovasc Dis 2019;112:135–43.
[5] Wang B, Prejean SP, Singh SP, *et al*. Percutaneous repair of Raghib syndrome. JACC Cardiovasc Interv 2020;13:e159–60.
[6] Garg A, Agrawal D, Mishra D, *et al*. Ostium primum atrial septal defect with persistent left superior vena cava opening into unroofed coronary sinus—a rare entity. Echocardiography 2019;36:1421–2.
[7] Shen JJ, Pan W. Prenatal diagnosis of a left superior vena cava draining into the left atrium and atrial septal defect with ventricular septal defect. Quant Imaging Med Surg 2023;13:553–7.
[8] Savu C, Petreanu C, Melinte A, *et al*. Persistent left superior vena cava—accidental finding. In Vivo 2020;34:935–41.
[9] Pérez-Caballero R, Plata Izquierdo B, Gil-Jaurena JM. Raghib syndrome. Surgical Treatment Rev Esp Cardiol (Engl Ed) 2016;69:71.
[10] Kumar B, Kodliwadmath A, Upadhyay AN, *et al*. Cor triatriatum with supramitral ring: "cor tetratriatum", associated with Raghib syndrome with Eisenmenger syndrome: multimodality imaging approach in this exceedingly rare case report. Egypt Heart J 2021;73:66.
[11] Okumori M, Hyuga M, Ogata S, *et al*. Raghib's syndrome: a report of two cases. Jpn J Surg 1982;12:356–61.
[12] Awasthy N, Ambatkar P, Radhakrishnan S, *et al*. Lutembacher syndrome with unroofed left superior vena cava: a diagnostic dilemma. Pediatr Cardiol 2013;34:1985–8.
[13] Nair VV, Rajashekar P, Saxena A, *et al*. Cortriatriatum with classical Raghib complex: a rare anatomic association. World J Pediatr Congenit Heart Surg 2014;5:318–20.
[14] Kawasuji M, Aoyama T, Kawajiri F, *et al*. [Surgical correction of Raghib's syndrome (termination of the left superior vena cava in the left atrium, atrial septal defect and absence of coronary sinus)]. Nihon Kyobu Geka Gakkai Zasshi. 1987;35:251–5.
[15] Bisbee CR, Sherard C, Rajab TK. Left superior vena cava draining to left atrium: a case report, review of the literature, and classification [published online ahead of print, 2023 Sep 2]. Pediatr Cardiol 2023;23:32–89. doi:10.1007/s00246-023-03289-5
[16] Sohrabi C, Mathew G, Maria N, *et al*. The SCARE 2023 guideline: updating consensus Surgical CAse REport (SCARE) guidelines. Int J Surg Lond Engl 2023;109:1136.