Scientific Research Publishing

World Journal of Cardiovascular Surgery, 2022, 12, 219-228
https://www.scirp.org/journal/wjcs
ISSN Online: 2164-3210
ISSN Print: 2164-3202

Laubry-Pezzi Syndrome in an Adult Male Patient and Its Surgical Correction: A Case Report and Literature Review

Md. Abir Tazim Chowdhury[1*], Md. Zulfiqur Haider[1], Sohail Ahmed[1], S. M. A. Zulker Nine[1], Arup Khan[1], Naheed Rayhan[1], Musfaqus Shalehin[1], Md. Sifat Uddin[1], Nakib Shah Alam[2], Saiful Islam Khan[2], Niaz Ahmed[2], Munama Magdum[3]

[1]Department of Cardiothoracic and Vascular Surgery, Evercare Hospital, Dhaka, Bangladesh
[2]Department of Cardiothoracic Anesthesia, Evercare Hospital, Dhaka, Bangladesh
[3]Department of Community Medicine, Shaheed Monsur Ali Medical College, Dhaka, Bangladesh
Email: *chowdhuryabir0@gmail.com

How to cite this paper: Chowdhury, Md.A.T., Haider, Md.Z., Ahmed, S., Zulker Nine, S.M.A., Khan, A., Rayhan, N., Shalehin, M., Uddin, Md.S., Alam, N.S., Khan, S.I., Ahmed, N. and Magdum, M. (2022) Laubry-Pezzi Syndrome in an Adult Male Patient and Its Surgical Correction: A Case Report and Literature Review *World Journal of Cardiovascular Surgery*, **12**, 219-228.
https://doi.org/10.4236/wjcs.2022.1210019

Received: August 25, 2022
Accepted: October 5, 2022
Published: October 8, 2022

Abstract

This Laubry-Pezzi syndrome is rarely seen in patients with ventricular septal defect (VSD) combined with aortic regurgitation. A 20-year-old male patient presented to the Cardiothoracic and vascular surgery department with a large VSD with severe aortic regurgitation (AR). However, he was first symptomatic at 12 years of age and initially treated by medical management with the suggestion of surgical correction. Nevertheless, he was delayed getting surgical management due to his financial problem. This article reported on an adult patient with Laubry-Pezzi syndrome and his surgical correction and outcome. The association of congenital defects VSD and AR needs to be identified and corrected in early life for better outcomes.

Keywords

Laubry-Pezzi syndrome, Adult Ventricular Septal Defect (VSD), Aortic Regurgitation (AR), Surgical Correction of Adult VSD

1. Introduction

Laubry-Pezzi syndrome is a combination of the ventricular septal defect (VSD) and aortic regurgitation (AR) due to prolapse of the right coronary or, less often, the non-coronary cusp [1], and it results from Venturi's effect on the aortic valve [2]. Early VSD closure has been suggested to contain the onset of Aortic Insuffi-

ciency (AI) or the worsening of the existent AR. Regardless, once an aortic valve disfigurement is present, surgical closure of isolated VSD without intervention to the aortic valve may not be sufficient to prevent the progression of AR. These patients need aortic valve repair (AVr) or aortic valve replacement (AVR). Aortic valve prolapse and AR are more frequent and severe in patients with delayed surgery, emphasizing the importance of early surgical intervention. It is a challenging surgical issue in patients with VSD and concomitant AR. In the treatment of AR, AVr has been an appealing alternative to AVR. Nevertheless, AVr is difficult for the elderly age group [3]. This case report describes a patient with Laubry-Pezzi syndrome and its surgical management.

2. Case Report

A 20-year-old male patient was admitted to our cardiothoracic and vascular surgery department as a diagnosed case of VSD with AR. At age 12, the patient's diagnosis was confirmed as large VSD with mild AR for the first time and advised for surgical correction. However, the patient's family could not bear the cost of his surgery, so they have avoided surgical management and taken symptomatic medical management to date. As a result of the delay, he deteriorated his physical condition gradually and presented to us with New York Heart Association (NYHA) functional dyspnea class III symptoms, palpitation, and chest pain. During the physical examination, his oxygen saturation was 98% in room air; pulse was 95/minute, sinus rhythm, high volume, water hammer nature, and wide pulse pressure with a systolic blood pressure of 140 mm of Hg and diastolic blood pressure of 40 mm of Hg. The patient had raised jugular venous pressure and the presence of bilateral ankle edema. Precordial examination demonstrated the following findings: visible precordial pulsation; apex beat shifted downwards and left lateral to the midclavicular line, at the sixth intercostal space (ICS), which was heaving in nature; palpable thrill on the left parasternal area at 3rd and 4th ICS, and also at 2nd ICS along the right parasternal area. A pansystolic murmur (grade 4/6), more prominent at 3rd and 4th ICS along the left parasternal area irradiating to the left axilla.

Furthermore, there was long aortic runoff, grade 2/6. Lung field auscultation revealed bilateral basal crepitant rales. Chest X-ray (**Figure 1**) showed an enlarged cardiac silhouette with prominent pulmonary vasculature suggestive of active congestion. An electrocardiogram (ECG) (**Figure 2**) demonstrated features of the typical Katz-Wachtel phenomenon suggestive of biventricular hypertrophy. Furthermore, transthoracic echocardiography (TTE) (**Figure 3**) demonstrates—a perimembranous restrictive VSD (9 mm at the left ventricular side and 5 mm at the right ventricular side) with a left to right shunt (PPG: 105 mm hg), aneurysmal dilatation and prolapse of RCS causing dynamic obstruction of the right ventricular outflow tract (RVOT), severe eccentric AR, left atrium (LA) and left ventricle (LV) hugely dilated Mild global hypokinesia of LV, and fair LV systolic function (Ejection fraction 50%).

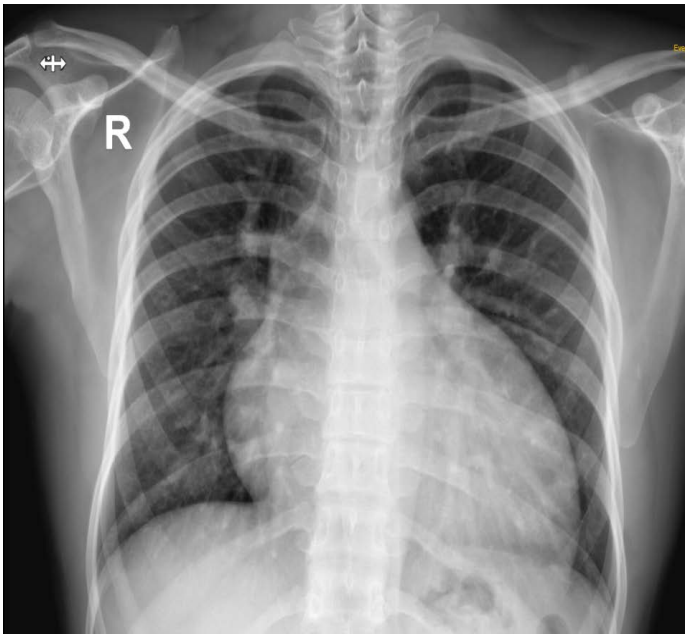


Figure 1. Preoperative chest X-ray demonstrates prominent pulmonary vasculature (active congestion) without pleural effusions. Enlarged cardiac silhouette.

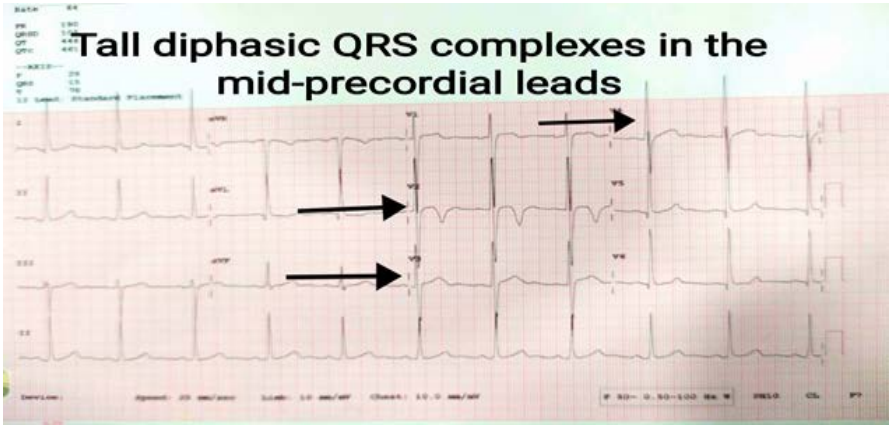
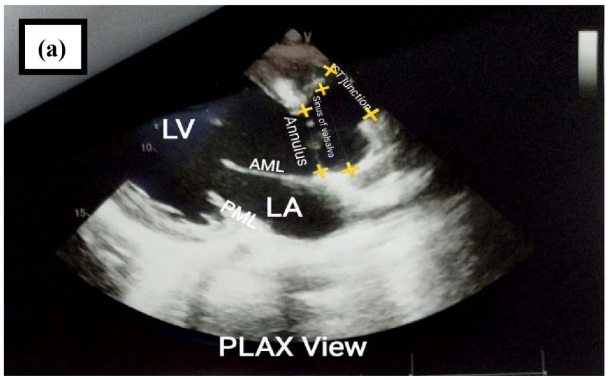


Figure 2. Preoperative Electrocardiogram shows typical Katz-Wachtel phenomenon *i.e.*, tall diphasic QRS complexes over 50 mm (black arrows) in either leads V2, V3, or V4. Suggestive of biventricular hypertrophy.



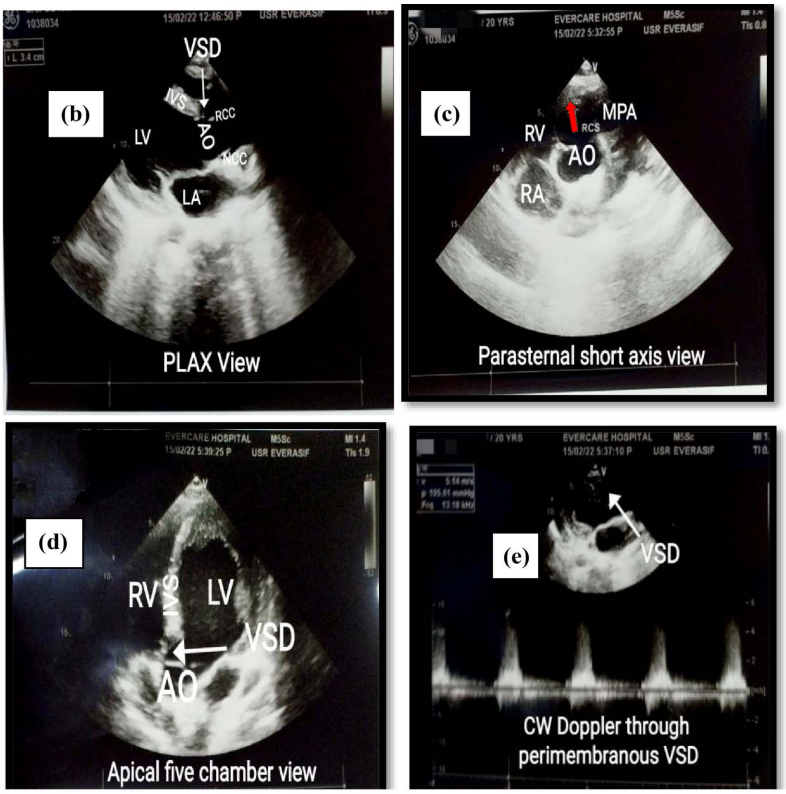


Figure 3. Images (a)-(e) represent preoperative transthoracic echocardiographic. (a) and (b) are parasternal long-axis (PLAX) view; (c) is para sternal short axis view, (d) is Apical five-chamber view; and (e) is continuous-wave Doppler evaluation through VSD. White arrows in the images identify the peri membranous ventricular septal defect (VSD), yellow marked areas in image 'A' demonstrate the measurement of the aortic root, and red arrow in the image 'C' indicating RCS prolapse to right ventricular out-flow. AO (Aorta), VSD (ventricular Septal Defect), RA (Right atrium), LA (Left atrium), LV (left ventricle), MPA (Major Pulmonary artery), RCC (Right coronary Cusp), RCS (Right Coronary Sinus), NCC (Non coronary Cusp).

The surgery was conducted under general anesthesia through a median sternotomy. The standard cardiopulmonary bypass (aortic bicaval cannulation) was established with aortic cross-clamping. The heart was arrested at diastole by a cardioplegic (CP) solution. Initially, delivery of the CP solution was through an aortic root cannula, and then an oblique aortotomy was performed that allows direct administration of the blood cardioplegia in the coronary Ostia (repeated every 10 - 15 minutes) and thus regurgitant aortic valve analysis. During the CPB maintenance, the body temperature was at a moderate hypothermia state. The heart was a hugely dilated, thickened wall, and there was no other external pathology noted. Through the transaortic exposure initial assessment of the aorta and VSD was conducted. Distorted aortic valve anatomy with dilatation of the valve annulus was noted, cusps tissues were unhealthily friable, and the right coronary sinus had aneurysmal changes. Five 5/0 stitches are placed on each commissure to visualize the aortic valve better. However, through the transaortic exposure, VSD was not adequately outlined; therefore, to expose VSD nicely, a

right atriotomy was performed. Through the right atriotomy approach, lifting the septal leaflet of the tricuspid valve (TV), a large subaortic-subpulmonary VSD (approx. 20 mm in diameter) was noticed, where the right aortic cusp was found prolapsed. Later, excision of the aortic valve's diseased cusps and valve sizing were performed through the transaortic route. The routes, as mentioned earlier, did not provide adequate visualization of the VSD outline, so we decided to put an incision at RVOT. After a fair determination of the VSD margin, we used polytetrafluoroethylenes (PTFE) patch for VSD closure, which was 20 mm in diameter. In the right coronary sinus area, horizontal mattress sutures with pledgetted 2-0 PTFE were passed from the crest of the interventricular septum and brought out through the aortic annulus, which on pulling up approximated the crest with the aortic annulus. In the other areas, everting annular sutures were taken. A few stitches were taken through the opening of the RVOT incision, thereby closing the VSD. And then, closure of the RVOT incision was performed. Later, the aortic valve was replaced with a 25 mm bi-leaflet mechanical valve with 2/0 ethibond interrupted stitching. The valve testing showed freely moving valve leaflets. The aortotomy incision was closed. Weaning from CPB was done slowly, and surgery was completed after ensuring adequate hemostasis. Figure 4 shows the operative techniques to correct the VSD defects and AVR.

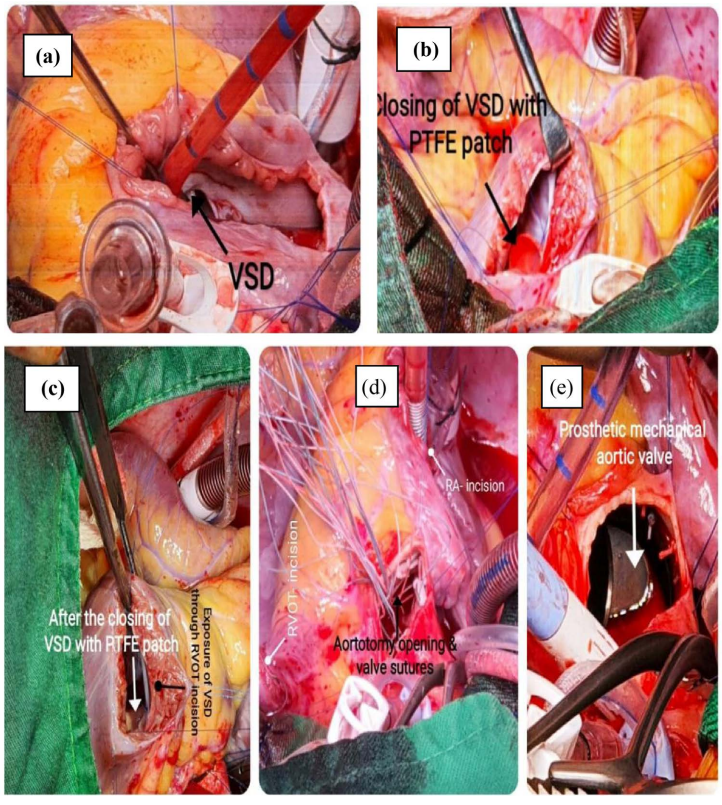


Figure 4. Images (a)-(e) represent operative correction techniques of VSD and AR. (a) showing VSD through RA incision; (b) showing VSD closure technique; (c) showing VSD closure with PTFE patch through RVOT approach; (d) showing valve suturing through aortotomy opening; (e) showing mechanical prosthetic valve placement on AV annulus.

The patient was kept in the cardiothoracic intensive care unit (CT-ICU) for 72 hours to monitor his hemodynamics, and the period was uneventful. A post-surgical bedside echocardiogram was performed, which demonstrated a prosthetic aortic valve and VSD patch in situ, aortic valve PPG 30 mm Hg with trivial central AR, and a VSD patch intact with a paradoxical inter-ventricular septum. Moreover, there was dilatation of left heart chambers, global hypokinesia, and EF (%) 40% noted. On the seventh postoperative day, the patient was discharged home with the following prescribed medication: TABLET WARFARIN 5 MG (once daily with maintained International Normalized Ratio chart, and target value was set 2 - 3), TABLET RAMIPRIL (2.5 MG once daily), TABLET METOPROLOL (25 MG once daily). At the 6-month follow-up visit, he improved his NYHA functional class (III to I), and a transthoracic echocardiogram (**Figure 5**) demonstrated a well-functioning prosthetic aortic valve. However, in situ VSD patch with small residual VSD (<2 mm) was noted with a left to right shunting. We continued the medication and made a schedule for follow-up with Echocardiogram one year after surgery and every six months for the following year.

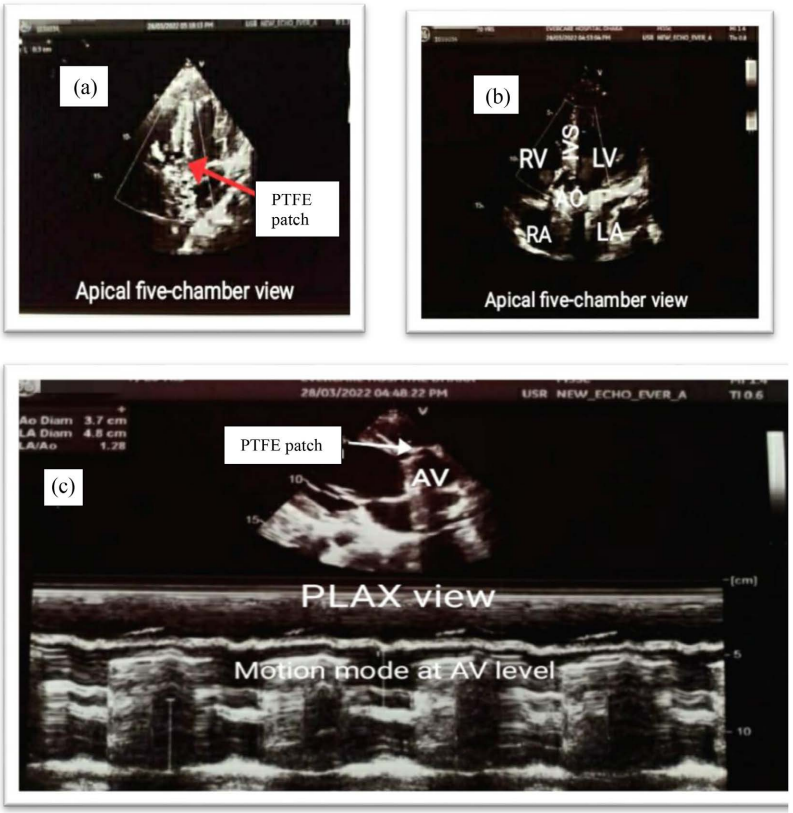


Figure 5. Images (a)-(c) represent postoperative transthoracic echocardiographic. (a) and (b) are apical five chamber view; (c) is parasternal long-axis (PLAX) view with motion mode at Aortic valve (AV) level. Red arrow in the image A, and white arrow in the image C identify the site PTFE patch closure of VSD. AV (Aortic valve), VSD (ventricular Septal Defect), RA (Right atrium), RV (Right ventricle), LA (Left atrium), LV (left ventricle), IVS (Inter Ventricular Septum).

3. Discussion

In 1921, Laubry and Pezzi first described a case of VSD with aortic insufficiency due to prolapsed aortic cusp. This anatomical and functional association typically appears in supracristal VSD (anterior and high defects), but it may present in perimembranous or muscular VSD. Such a defect causes prolapse of the aortic right coronary cusp due to both scarcity of leaflet support and Venturi's phenomenon [2]. The pathophysiology of the Venturi's effect in the case of VSD is described as when blood traverses through the defect and if it is situated beneath the aortic valve where the fall of pressure aspirates the right coronary or non-coronary cusp resulting in AI or AR [4]. AI complicates the course of patients' subcristal VSDs five times as often as patients with perimembranous VSDs [5]. Understanding of the natural history of Laubry-Pezzi syndrome is little in the literature. Symptomatic presentation of VSD in earlier age groups, and recommendation for early correction of VSD and AI, might be the reason behind this [6]. Laubry-Pezzi syndrome involves aortic leaflet prolapse, aortic cusp retraction, aortic annulus dilatation, and deformation of the adjoining sinus of Valsalva. Damages to the aortic valve are irreversible, and AR will keep on deteriorating without surgery. Advancement to severe AR leads to AVr or AVR or even Ross or Bentall procedures when the sinuses are abnormal. These patients show an increased chance of infectious endocarditis and progressive aortic valve malfunction. Earlier surgical repair is indicated as soon as aortic regurgitation ensues [7]. In the present case, the patient's age when he was first diagnosed with a congenital heart defect was 12 years old, and he was seeking surgical correction at 20 years. Regarding the age of the detection of AR, detection trends are mainly between the ages of 3 - 8 years [7]. Furthermore, the literature reported that elderly patients with such kind of defect presented with more severe AR [3]. The findings of aortic pathology in the current case correspond to several pieces of literature that mentioned that, in the later stages of the disease presentation due to structural abnormalities in the sinus and the ascending aorta, aneurysmal dilatation of the aortic root and ascending aorta is possible [2] [8] [9]. Moreover, in the current case, a defect in the ventricular septum with echocardiographic imaging was a large perimembranous type, and in the earlier text, it was mentioned that this variant of VSD was less complicating than sub arterial one to forcing AI [5]. Echocardiography is an essential diagnostic tool for detecting structural anomalies of the heart, but it is unreliable in providing the actual size of the VSD, especially in the presence of AR, because the proper size tends to be underestimated. The best method would still be the intraoperative measurement of the VSD defect [3], which was performed in this case and found a disparity between echocardiographic measurements. Furthermore, per-operatively, VSD was located sub arterial (subaortic-subpulmonary). In this case, we did not perform other diagnostic investigation tools like a Transesophageal echocardiogram, heart catheterization, computerized cardiac tomography, or magnetic resonance imaging to evaluate the case further. The decision was made due to our

clinical, electrocardiographic, and echocardiographic assessments. The lack of financial solvency of the patient to carry out those tests was also an important reason behind it. However, we discussed the importance of these investigations with the patient. In the current case, the hemodynamic effects of VSD can cause biventricular enlargement, which was demonstrated in the ECG. Interestingly, it is typical of Katz-Wachtel phenomenon, and it is described as the presence of tall diphasic QRS complexes at least 50 mm in altitude in lead V2, V3, or V4-mid precordial leads [10] and large anterior electrocardiographic forces are due to the hypertrophied right ventricle (RV) and late posterior forces due to the hypertrophied LV [11]. Surgical treatment of Laubry-Pezzi syndrome is usually satisfactory [12], and the trans-aortic approach usually allows a single-step procedure to repair the septal and the aortic disorders simultaneously [13]. However, due to the complexity of the defects, we were approached through RA and RVOT in addition to aortotomy. In this reported case, we closed the VSD with a large PTFE patch, and our technique was more or less to Yacoub's technique [14]. The Avr may be recommended when attainable to evade lifetime anticoagulant therapy for such young patients [15]. Nevertheless, it was not feasible in the current case due to gross structural changes in the aortic root, so we replaced the patient's AV with a mechanical prosthetic valve [2] [3]. The initial repair consisted of patients treated for VSD and AR needing a close long-term follow-up of VSD closure, the aortic valve, and the aortic root, specifically when preoperative AR was moderate to severe and when the surgery occurred at an older age. In the reported case, we followed the patient six months after his surgery and found a well-functioning prosthetic valve with a VSD patch in situ with one residual VSD (<2 mm). Very few reports deal with long-term outcomes, complications, and reoperation rates. In this reported case, we found a residual VSD, a classical complication (not requiring a reoperation systematically) after surgical closure of a VSD [16]. Although, in our initial postoperative echocardiogram, it was absent. However, another complication that could be encountered in such corrective surgery is aortic root pseudoaneurysm, and such complication needs a redo surgery in most cases [16]. We are fortunate that we did not encounter such aortic complication, although the current patient's aortic root had aneurysmal dilation prior to surgery.

The report and knowledge of surgical corrections and complications in patients with Laubry-Pezzi syndrome are of utmost priority for several reasons. In western countries, around 85% of children with congenital heart disease (CHD) endure into adulthood [17], and the prevalence of adults with CHD is steadily growing in developed countries. Interestingly, it now outnumbers the prevalence of children with CHD [18]. From the discussion mentioned above, our recommendations are:

1) Early recognition and correction of such rare CHD to prevent complications at a later age.

2) Report such cases more frequently whenever they are encountered to

enrich the literature.

3) Report all the potential disease complications and surgical techniques for their correction to enrich the knowledge and awareness among cardiac surgeon's community and the patients.

4. Conclusion

The Laubry-Pezzi syndrome is a rare congenital heart disease ushering to a substantial alteration of cardiac hemodynamics. Screening in early life with echocardiography is essential for the diagnosis; however, it does not always accurately measure the defect. The mainstay of the correction of this defect is surgery. The surgical correction of VSD and AR at an early age is vital to preserving the aortic valve function and preventing more radical surgery for aortic pathology.

Ethics Approval and Informed Consent

This article does not contain any studies involving human participants or animals conducted by the author. Informed consent was obtained from the patient in the case report.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Sbizzera, M., Pozzi, M., Cosset, B., Koffel, C., *et al*. (2016) Long-Term Complications after Surgical Correction of Laubry-Pezzi Syndrome. *Journal of Thoracic Disease*, **8**, E232. https://doi.org/10.21037/jtd.2016.02.28

[2] Piazza, F., Santoro, G. and Russo, M.G. (2013) Aortic Insufficiency Due to Ventricular Septal Defect (Laubry-Pezzi Syndrome). *Journal of Cardiovascular Medicine*, **14**, 164-165. https://doi.org/10.2459/JCM.0b013e3283515c30

[3] Krishnasamy, S., Sivalingam, S., Dillon, J., *et al*. (2021) Syndrome of Ventricular Septal Defect and Aortic Regurgitation—A 22-Year Review of Its Management. *Brazilian Journal of Cardiovascular Surgery*, **36**, 807-816. https://doi.org/10.21470/1678-9741-2020-0207

[4] Bhat, A.N., Sallehuddin, A., Riyas, M., *et al*. (2013) Further Insights into the Syndrome of Prolapsing Non-Coronary Aortic Cusp and Ventricular Septal Defect. *Global Cardiology Science and Practice*, **2013**, 91-95. https://doi.org/10.5339/gcsp.2013.11

[5] Tweddell, J.S., Pelech, A.N. and Frommelt, P.C. (2006) Ventricular Septal Defect and Aortic Valve Regurgitation: Pathophysiology and Indications for Surgery. *Seminars in Thoracic and Cardiovascular Surgery*: *Pediatric Cardiac Surgery Annual*, **9**, 147-152. https://doi.org/10.1053/j.pcsu.2006.02.020

[6] Radhakrishnan, B.K., Idhrees, M.., Devarajan, S., *et al*. (2014) Primary Modified Bentall's Procedure in a Case of Laubry-Pezzi Syndrome. *The Annals of Thoracic Surgery*, 98, 1445-1447. https://doi.org/10.1016/j.athoracsur.2013.11.070

[7] Tweddell, J.S., Pelech, A.N., Jaquiss, R.D., *et al*. (2005) Aortic Valve Repair. *Semi-*

nars in Thoracic and Cardiovascular Surgery: *Pediatric Cardiac Surgery Annual*, **2005**, 112-121. https://doi.org/10.1053/j.pcsu.2005.01.013

[8] Martínez-Quintana, E., Rodríguez-González, F. and López-Gude, M.J. (2013) Laubry-Pezzi Syndrome with Aortic Root Dilatation Treated with a Bentall and De Bono Procedure. *World Journal for Pediatric and Congenital Heart Surgery*, **4**, 299-301. https://doi.org/10.1177/2150135113484160

[9] Sakakibara, S. and Konno, S. (1962) Congenital Aneurysm of the Sinus of Valsalva Anatomy and Classification. *American Heart Journal*, **63**, 405-424. https://doi.org/10.1016/0002-8703(62)90287-9

[10] Elliott, L.P., Anderson, R.C., Tuna, N., Adamsjr, P.A. and Neufeld, H.N. (1963) Complete Transposition of the Great Vessels: II. An Electrocardiographic Analysis. *Circulation*, **27**, 1118-1127. https://doi.org/10.1161/01.CIR.27.6.1118

[11] Varriale, P.H., Kennedy, R.J. and Alfenito, J.C. (1969) Vectorcardiogram of Combined Ventricular Hypertrophy: Posterior Counterclockwise Loop (Frank System). *British Heart Journal*, **31**, 457. https://doi.org/10.1136/hrt.31.4.457

[12] Pontailler, M., Gaudin, R., Moreau de Bellaing, A. and Raisky, O. (2019) Surgical Repair of Concomitant Ventricular Septal Defect and Aortic Cusp Prolapse or Aortic Regurgitation, also Known as the Laubry-Pezzi Syndrome. *Annals of Cardiothoracic Surger*, **8**, 438-440. https://doi.org/10.21037/acs.2019.05.11

[13] Riache, A., Kebour, D. and Sayah, T. (2020) Surgical Repair of Perimembranous Ventricular Septal Defect and Aortic Regurgitation in an Adult Patient with Laubry-Pezzi Syndrome. *Junior Medical Research*, **3**, 15-17. https://doi.org/10.32512/jmr.3.1.2020/15.17

[14] Yacoub, M.H., Khan, H., Stavri, G., Shinebourne, E. and Radley-Smith, R. (1997) Anatomic Correction of the Syndrome of Prolapsing Right Coronary Aortic Cusp, Dilatation of the Sinus of Valsalva, and Ventricular Septal Defect. *The Journal of Thoracic and Cardiovascular Surgery*, **113**, 253-261. https://doi.org/10.1016/S0022-5223(97)70321-0

[15] Sbizzera, M., Pozzi, M., Cosset, B., Koffel, C., *et al*. (2016) Long-Term Complications after Surgical Correction of Laubry-Pezzi Syndrome. *Journal of Thoracic Disease*, **8**, E232-E234. https://doi.org/10.21037/jtd.2016.02.28

[16] Quevedo, H.C., Santiago-Trinidad, R., Castellanos, J., *et al*. (2014) Systematic Review of Interventions to Repair Ascending Aortic Pseudoaneurysms. *Ochsner Journal*, **14**, 576-585.

[17] Khairy, P., Ionescu-Ittu, R., Mackie, A.S., *et al*. (2010) Changing Mortality in Congenital Heart Disease. *Journal of the American College of Cardiology*, **56**, 1149-1157. https://doi.org/10.1016/j.jacc.2010.03.085

[18] Webb, G., Mulder, B.J., Aboulhosn, J., *et al*. (2015) The Care of Adults with Congenital Heart Disease across the Globe: Current Assessment and Future Perspective: A Position Statement from the International Society for Adult Congenital Heart Disease (ISACHD). *International Journal of Cardiology*, **195**, 326-333. https://doi.org/10.1016/j.ijcard.2015.04.230