

# Single-stage Procedure of Coarctation Aorta and Pectus Excavatum Repair: a Rare Case Report

Christian I.H. Gulo<sup>1,\*</sup>, Arief Rahman Hakim<sup>1</sup>, Heroe Soebroto<sup>2</sup>, Erdyanto Akbar<sup>2</sup>

<sup>1</sup>Resident of Cardiothoracic and Vascular Surgery, Faculty of Medicine, Universitas Airlangga, Surabaya, Indonesia

<sup>2</sup>Cardiothoracic and Vascular Surgeon, Faculty of Medicine, Universitas Airlangga, Surabaya, Indonesia

Corresponding author: medisch\_126@yahoo.com

## ABSTRACT

Coarctatio of aorta (CoA) is one of congenital heart anomaly which is defined as narrowing the aorta, especially descending aorta and commonly happen in male newborn. This anomaly usually associated with other abnormalities of cardiac. Another abnormality which more common in children is pectus excavatum. The combination of these is rare and the strategy of operation should be considered wisely. Case Presentation: A 8-year-old girl patient was brought by her mother to public health centre because of vomiting and low intake. Her examination revealed that she had problem with his heart and referred to Dr. Soetomo Hospital. In the hospital, she was diagnosed by coarctatio aorta and pectus excavatum from the examination. Then, she underwent surgery with single-stage procedure and the result was satisfactory without any complication. Discussion : The combination of Coarctatio of aorta and pectus excavatum is rare. Both of them have to be done by surgery. There is no guideline or consensus dictate which one must be done at first. There are some arguments about the procedure. But single-stage procedure can be considered as first option of treatment. Conclusion: Combination procedure to repair coarctatio aorta (CoA) and pectus excavatum in single-stage operation procedure can be done safely and the result is satisfied. But it will need further research to establish this procedure.

**Keywords:** coarctatio of aorta, pectus excavatum, open surgical repair

## 1. INTRODUCTION

Coarctatio of aorta (CoA) is one of type of congenital heart anomaly which approximately happen in 3 cases per 1000 births or 4-6 percent of live birth with congenital heart disease.[1] CoA is defined as discrete narrowing of aorta, especially Descending aorta with various anatomical site along it. The classic CoA location is in the distal of Left Subclavian Artery or post ductal or proximal to ductus arteriosus (pre-ductal) and usually associated with other congenital heart diseases, for example bicuspid aortic valve (BAV) for most common (45%-60%), ventricular septal defect (VSD), patent ductus

arteriosus (PDA) and transposition of great artery (TGA) [2]. The degree of narrowing is more important to clinical relevance and the surgery is the treatment of choice, either be done by open surgery or endovascular surgery. Another congenital abnormality which commonly happen is Pectus Excavatum with almost 1:3-400 in newborn, especially in male newborn [3]. The diagnosis of Pectus Excavatum is mainly determined by clinical examination. It can be identified during infancy but more prominent during and after puberty and also its treatment is surgery with various procedures [4]. CoA patient with pectus excavatum is rare combination

and the strategy to their surgery must be considered wisely. In this paper, we will present the case and the treatment that we have done in our hospital.

## 2. CASE PRESENTATION

A girl 8-year-old patient was brought by his mother to the public health centre complaining with vomit and low intake and then her body was getting thinner. Then, she referred to the HVA Hospital in Pare Kediri. Echocardiography was performed and resulted a defect of her cardiac. Furthermore, the patient referred to Dr. Soetomo hospital in Surabaya to check her abnormality. In Dr. Soetomo Hospital, patient underwent physical exam, chest x-ray, echocardiography and diagnostic cardiac catheterization. From physical examination, vital sign was normal and the differentiation of systolic blood pressure between upper and lower extremity more than 20 mmHg (112/83 in upper extremity and 85/54 in lower extremity). Her chest wall was abnormal, called pectus excavatum, and from auscultation, a systolic murmur was found at ICS II – IV Left Parasternal line without gallop. Chest x-ray was normal (Fig.1). Then, from the echocardiography, it was found that the solitus site, AV concordance, pulmonary venous drainage, and heart chambers were normal but there were mild AR subaortic stenosis and coarctatio of aorta (Fig.2). On cardiac catheterization, the coarctatio aorta could be seen better, located at descending aorta, distal to Left Subclavian Artery (Fig.3). After all the examinations had been performed, patient was planned to have a single-stage surgery to correct the coarctatio aorta as well as pectus excavatum.



figure 1. Chest X-ray before the operation

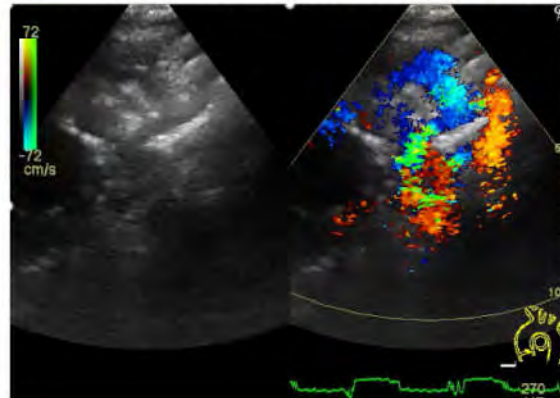


figure 2. Echocardiography from suprasternal view. There was turbulating in the descending aorta, distal to Left Subclavian Artery

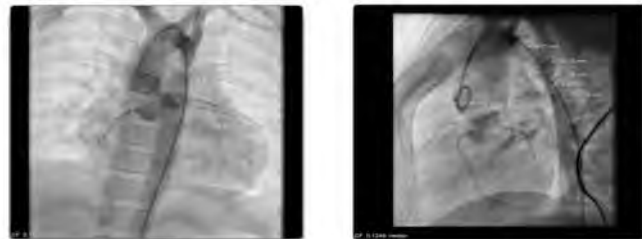


figure 3. Aortography. There was coarctatio aorta distal to Left



figure 4. Pectus Excavatum Pre Procedure Intraoperative, first this patient was performed with a median sternotomy and found

pectus excavatum, a large heart with normal contractility, juxta ductal aortic coarctatio, and large PLSVC that closed the aortic coarctatio. PDA ligation was performed. Because the exposed of descending aorta was blocked by PLSVC, it was decided to perform a left posterolateral thoracotomy. On the left posterolateral thoracotomy, a juxta ductal coarctation crossed over the descending aorta. Then performed aortic coarctatio resection with end-to-end anastomosis. After that, second median sternotomy was performed and the aortic valve was evaluated and the subaortic ridge was found. Then, a subaortic ridge was resected and pectus excavatum repair were performed with a modified pectus bar. After surgery, postoperative transesophageal echocardiography (TEE) was performed, and obtained qualitative mild AR, without jet and subaortic membrane and good descending aortic flow.

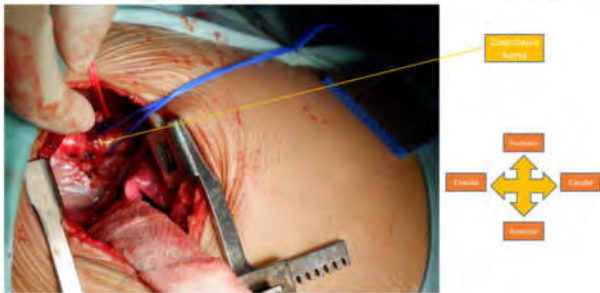


Fig.5. Juxta Ductal Coarctation Aorta before repair

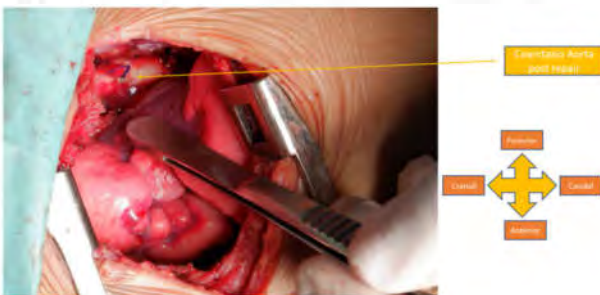


Fig.6. Coarctation Aorta after repair

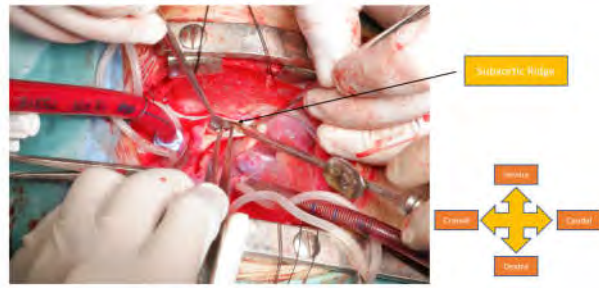


Fig. 7. Subaortic ridge resection

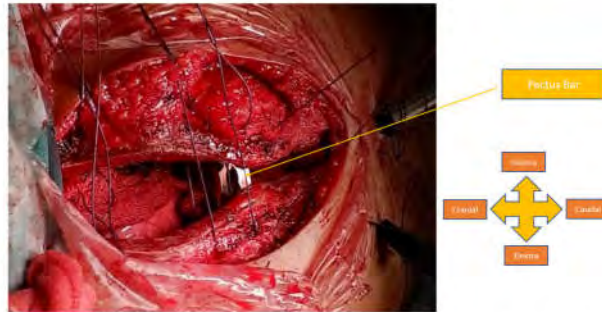


Fig. 8. Pectus Excavatum repair with Pectus Bar

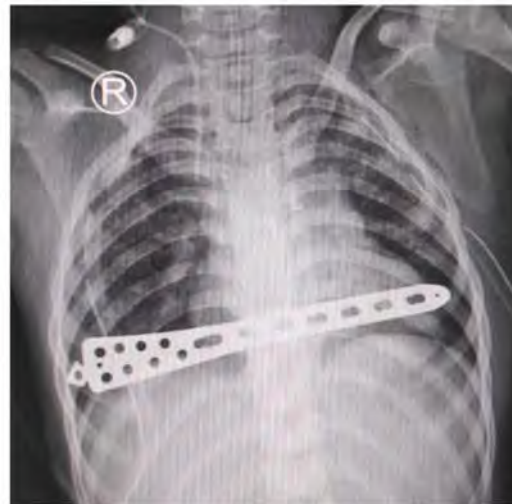


Figure 9. Chest X-ray



## 1. DISCUSSION

Coarctatio of aorta (CoA) is one of type of congenital abnormality, presented by narrowing lumen of the aorta, especially descending aorta [5]. There are various ana-

tomical sites of CoA and it implicates the clinical presentation. Because of narrowing of the aortic lumen, then it increases left ventricular afterload and reduced perfusion of lower extremity resulting upper extremity hypertension due to activation of Renin-Angiotensin Aldosterone system in the renal [2]. From this case, the systolic blood pressure in upper extremity is higher than lower extremity, the differentiation between them is more than 20mmHg.

Beside upper extremity hypertension, other compensatory mechanisms are concentric left ventricular hypertrophy, development of collateral flow and post stenotic dilation as seen in this case. The indication of CoA intervention based on several things, such as Non-invasive systolic blood pressure gradient of >20 mmHg between upper and lower limbs, Peak-to-peak transcatheter gradient of  $\geq 20$  mmHg across the coarctation site, Peak-to-peak transcatheter gradient of <20 mmHg in the setting of extensive collateral circulation around the coarctation site, significant left ventricular hypertrophy, uncontrolled systemic hypertension, and abnormal blood pressure during exercise stress test [6]. The treatment of CoA can be divided into two, open surgical repair, including end-to-end anastomosis, graft interposition, patch angioplasty, or subclavian flap and endovascular repair with balloon angioplasty or stenting [1]. The choice of treatment depends on age, complexity of coarctation, and other cardiac or extracardiac defect. According to this case, beside CoA, patient also has funnel or sunken chest, called pectus excavatum, one type of anterior chest wall abnormality. The treatment of pectus excavatum done by surgery, either Ravitch or Nuss procedure. Nuss procedure by inserting metal bar using thoracoscopy, become more popular than Ravitch procedure because of minimally invasive and less compli-

cation after procedure [4]

The treatment of coarctatio aorta with pectus excavatum become more challenging because there are two procedures that will be performed and also increase the risk of morbidity and mortality to the patient. There is no guideline or consensus which focus on surgical approach to this case. Some authors suggest two-stage surgical procedure to avoid a complication of prolong duration of operation, but others suggest that single-stage procedure which is performed simultaneously achieves good result and minimal complication [7]. In this case, we performed single-stage operation procedure, beginning with repair of coarctatio aorta via left posterolateral thoracotomy and after that, via sternotomy to repair subaortic stenotic, including use of cardiopulmonary bypass machine as well as repair pectus excavatum by using modified pectus bar. The result was satisfactory without any complication.

## 2. CONCLUSION

Combination procedure to repair coarctatio aorta (CoA) and pectus excavatum in single-stage operation procedure can be done safely and the result is satisfied. But it will need further research to establish this procedure.

## REFERENCES

1. H. Suradi and Z. M. Hijazi, "Current management of coarctation of the aorta," *Glob. Cardiol. Sci. Pract.*, vol. 2015, no. 4, p. 44, 2015, doi: 10.5339/gcsp.2015.44.
2. P. Agasthi et al., "Management of adults with coarctation of aorta," *World J Cardiol*, vol. 12, no. 5, pp. 167–191, 2020.
3. A. S. Media, H. K. Pilegaard, and F. V. de Paoli, "Combining Correction of Pectus Excavatum and Open Heart Surgery in

- a Single-Stage Procedure," *Ann. Thorac. Surg.*, vol. 109, no. 1, pp. e71–e74, 2020, doi: 10.1016/j.athoracsur.2019.07.043.
4. A. Farronato et al., "Pectus excavatum in adolescents and children: The Nuss technique," *Pediatr. Med.*, vol. 2, no. 4, pp. 6–13, 2019, doi: 10.21037/pm.2019.07.02.
  5. R. D. Torok, "Coarctation of the aorta: Management from infancy to adulthood," *World J. Cardiol.*, vol. 7, no. 11, p. 765, 2015, doi: 10.4330/wjc.v7.i11.765.
  6. A. R. Doshi and S. Chikkabyrappa, "Coarctation of Aorta in Children," *Cureus*, vol. 10, no. 12, pp. 1–10, 2018, doi: 10.7759/cureus.3690.
  7. Y. Sun, P. Zhu, and S.-Y. Zheng, "Simultaneous repair of pectus excavatum and congenital heart disease," *J. Cardiothorac. Surg.*, vol. 9, no. 168, pp. 1–3, 2014, doi: 10.1186/s13019-014-0168-7.