Case Report

Berry syndrome; a successful one-stage repair in neonate periods, evaluation result after 9 years, a case report

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ABSTRACT

Background: IAA with an intact ventricular septum is distinctly unusual. Combination with an Aortopulmonary Window (APW), ascending aortic origin of the right pulmonary artery and PDA may be present which is called as Berry syndrome, a rare combination of cardiac anomalies, reported to be 0.046%, lethal combination and die shortly after birth.

Case Report: We report a 9 days-old male neonates weighing 3.85 kg was referred by local hospital to our center and was ventilated with history of respiratory distress and severe infection since he was born. Admitted to our PICICU, 2D echo showed an IAA type A associated with a huge APW type II and restrictif PDA. A PGE1 infusion was started, during the following days the baby experienced several epileptic episodes. After improvement of the clinical condition, surgery was performed on the 20th days of life on year 2011. A successful one-stage repair of such anomalies in which cutting of PDA that arised from PA trunk and distally becoming into descending aorta, extended end to end anastomosis to conduct the ascending aortic blood flow into the descending aorta and intra arterial baffle was used. A 4-0 Gore-Tex baffle was used both to close the APW and separated the RPA from aortic origin with a good result, as his recently grown up as a cheerful 9 year old child who is growing actively and has entered elementary school in grade 2.

Conclusion: Berry syndrome is a rare but well-identified and surgically correctable anomaly. Early diagnosis and surgical treatment to avoid irreversible pulmonary hypertension is mandatory.

1. Background

An interrupted aortic arch (IAA) is usually associated with a ventricular septal defect and a patent duct. The coexistence of an IAA with an intact ventricular septum is distinctly unusual. In such cases, an aortopulmonary window (APW), anomalous origin of the right pulmonary artery (RPA) arising from the ascending aorta is a rarely reported association of congenital heart anomalies [1–4], first described as a syndrome by Berry only in 1992 [1]. The incidence of this combination of anomalies among patients with congenital heart disease is reported to be 0.046% [5,6] with high mortality rate (90%) in the neonatal period, lethal combination; most affected infants die shortly after birth [7] and the surviving patients have development of pulmonary hypertension in most cases [4,8].

Berry et al. recognized the association of the distal type of aortopulmonary window with aortic origin of the right pulmonary artery and hypoplasia or interruption of the aortic arch as a specific syndrome. Correct preoperative diagnosis and detailed anatomic depiction of each component of this association are important [7,9].

Since the first case was reported by Berry, most repairs were performed in two-stage operations, but one-stage repair has been reported recently [10]. One-stage repair in the neonatal period immediately after the correct diagnosis is mandatory. For critically ill patients, decreasing the circulatory arrest and cardiac ischemic time is important [11]. In the English literature, this anomaly has been reported in nearly 100 patients. We also experienced this rare case, and successful one-stage repair was done in a 20-days-old neonate using an alternative technique without circulatory arrest. Written informed consent from his mother has given to publish this rare case. This case report has adjusted the content to SCARE 2020 criteria [12].

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2. Case report

A 9 days-old male neonates weighing 3.60 kg was referred by local hospital to our center (National Cardiac Center Harapan Kita Hospital/NCCHK) and was ventilated because of severe metabolic and respiratory acidosis, circulatory dynamics instability with history of respiratory distress, cyanosis, hyperbilirubinemia and severe infection that present since he was born on year 2011. The perinatal history was unremarkable and at a previous hospital she was born at 40 weeks of gestation by normal labour with a birth weight of 3.85 kg, oxygen saturation 90% and an APGAR score of 9/9. On the 6th day postpartum he presented with feeding difficulty, severe cyanosis and congestive heart failure and was referred to local hospital for further treatment, was ventilated at the emergency room with oxygen saturation 82%, and then transferred to us for possible surgical repair. Stable hemodynamic with oxygen saturation was 75%, while differential oxygen saturation was noted between the both upper and lower limbs (89%; 76% and 97%; 83%, respectively). No apparent cardiac murmur was detected. His chest X-ray showed mild cardiomegaly and congestion of the lungs, and pulmonary edema. Electrocardiogram showed normal rhythm, right axes

![Fig. 1. Long Axis view revealed a defect at AORPA (Arrow) from 2D Echocardiography and colour Doppler. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)](image1)

![Fig. 2. A. Apical view with tilting to the anterior view revealed a 10 mm defect of APW (Arrow), B. Supra sternal view showed Aortic arch with 3 branches (upper) and no communication with AoD. AoD arise from PDA (lower).](image2)

![Fig. 3. Preoperative situs demonstrating the anomalous origin of the RPA from the ascending aorta, AP Window type II and the IAA type A. PDA becoming into descending aortic Intra Operative Findings (arrows):A. AP Window type II B. Ascending Aortic origin of RPA C. Huge PDA that arise from PA Trunk and then becoming into descending aortic, confirmed the IAA type A with no communication after the 3rd arch branch.](image3)
deviation, and mild right ventricular hypertrophy.

Admitted to our pediatric cardiac intensive care unit (PCICU) with oxygen saturation between 40% and 70%. Two-dimensional echocardiography revealed an IAA type A and APW type II with a restricted PDA, pulmonary hypertension but no ventricular septal defect (Figs. 1 and 2). A PGE1 infusion 10 nano was started to re-open the ductus arteriosus and diuretics were administered. During the following days the baby had severe infection and DIC with increased of leucocyte 28.387, pro-calcitonine 11, D-dimer 4.800. Also had experienced of several epileptic episodes, and a brain sonography showed arachnoiditis, brain edema and no intracranial bleeding. Because of the risk of infection, sepsis and taking into account the good general condition, operation was deferred.

After improvement of the clinical condition, operation was performed on the 20th days of life. A one-stage surgical correction was performed through a median sternotomy under profound hypothermia (26 °C). Before the administration of heparin, the entire ascending aorta and the arch vessels, pulmonary trunk and both branch were mobilized.

Fig. 4. (A). Schematic drawing of intra operative findings and (B) operative surgical correction of Berry syndrome.

Fig. 5. Intra operatif findings: A (Arrow) shows 4-0 mm PTFE graft sewn to the right innominate artery for aortic cannulation, B (Arrow) the site of the future aortic arch-descending aortic anastomosis, C (Arrow) internal defect of AP window and aortic origin of RPA, D (Arrow) A 4-0 Gore-tex patch was used both to close the aortopulmonary window and separated the right pulmonary artery from aortic origin.
The external anatomy confirmed an IAA type A and a large APW type II, and this time the right pulmonary artery (RPA) was recognized to originate from the posterolateral part of the ascending aorta, as well as connection with common pulmonary artery. Aorta and pulmonary arteries (PA) were obvious and intact at their origin. The ascending aorta is then come to end by giving three branches including, brachiocephalic artery, left common carotid artery, and left subclavian artery. There was not evidence of descending aorta. Proximal part of PDA arose from PA trunk which is distally becoming into descending aorta with left pulmonary artery are origin from PA trunk also detected at the same time (Figs. 3 and 4).

Cardiopulmonary bypass (CPB) was established by the right hemicerebral perfusion and the bicaval drainage. The right hemicerebral perfusion was obtained via expanded 4-0 mm polytetrafluoroethylene (ePTFE) graft sewn to the right brachiocephalic artery. The aortic cross clamp was applied (one-third flow) and cardioplegia was infused. Aortic arch vessels were clamped at the origin of the brachiocephalic artery, the left common carotid artery, and left subclavian artery. Immediately there after, snaring of both pulmonary arteries precluded overloading the lungs, and the pump flow was lowered to 10% to perfuse only the ascending aorta was transversely incised distal to the AP window. In myocardium. Ductal tissue was ligated near its pulmonary origin and resected until normal aortic tissue appeared, then the ascending aorta function, no AP window residual.

IAA is characterised by a lack of luminal continuity between the ascending and descending aorta. It represents a critical ductus dependent congenital heart disease [16]. The association between IAA and APW and aortic origin of the RPA are rare, complex and represents a life-threatening malformation in the neonatal period [8,17], which poses challenging problems to both the cardiologist and the cardiac surgeon. Many forms were described according to the level of the aorto-pulmonary window and the location of the origin of the RPA. This malformation was found to be an intermediate type of truncus arteriosus, excepting that two different annulus are present [1,18,19].

Systemic circulation is ductus-dependent in this malformation, so it is very important to improve the metabolic status of these neonates before the operation with infusion of PGE1, and careful manipulation of artificial ventilation, if necessary, to improve systemic perfusion and prevent renal failure and low output syndrome [8]. Usually, the neonate presenting with such a complex anomaly is critically ill and may not tolerate any invasive examination; therefore a most complete diagnosis should be made by echocardiography. However, echocardiography did not allow the detection of the abnormal RPA origin in this case. Since this was recognized immediately after we opening the pericardium and did not increase the difficulty of the operation [19].

Satisfactory repair can be regarded as such only when it results in an unobstructed left ventricular outflow tract, aortic arch, RPA [20–22] and to restore a normal perfusion of the lower part of the body and to prevent the development of pulmonary damage [17,20]. Ideally, the growth potential of the aorta and the pulmonary artery should be maintained. The results of the one-stage repair of interrupted aortic arch and associated cardiac anomalies are relatively better than those of a two-stage approach in terms of both mortality and morbidity [11,17,19].

Surgical management has to take into account the age of the patient, the size and location of the APW, and the presence and severity of pulmonary hypertension. To correct this malformation many surgical solutions have been proposed, and there is still discussion on how close the APW (ligature, closure with a patch through the aorta or through the pulmonary artery) and how to correct the IAA [8,20]. We think that one-stage repair in the neonatal period is mandatory as soon as possible after the correct diagnosis, because the natural history of this malformation presents high early mortality and high risk of pulmonary hypertension, as previously documented in the literature [4,8,9,16].

Surgical correction of this complex anomaly requires both repair of the hypoplastic or interrupted aortic arch and septation of the APW, with exclusion of the RPA orifice from the aorta [2]. The success of this complicated repair depends on the meticulous design of each step of the procedure before surgery [9]. Most reports of the surgical correction of IAA in combination with other lesions, including AP window, have described operation on infants rather than neonates, and circulatory arrest has been used for repair. Closure of the AP window is best performed after completion of the arch repair [20–22].

From a technical point of view, the best surgical option is aortic arch repair by direct anastomosis, without the use of a prosthetic conduit, as described by Vouhe and associates [22,23] in patients with IAA and ventricular septal defect. The APW was closed with a pericardial or Gore-Tex patch and from the pulmonary artery to avoid damage and distortion of the aortic valve. The operative risk using this technique is the same or probably less than for patients with IAA and ventricular septal defect, and the midterm results are very satisfactory [1,8,11].

In our patient, we used hypothermic bypass at low flow without circulatory arrest. Complete repair of IAA has been achieved without the use of any synthetic material, by end to end anastomosis thus allowing the potential for future growth, but for APW, and anomalous origin of the RPA we use a synthetic material (Gore-Tex) while avoiding circumferential suture lines on all the anastomoses. The posterior wall of the ascending aorta was used to form the posterior portion of the confluence between the RPA and the main pulmonary artery, and a patch 4-0 gore-tex was used to constitute its anterior half, a technique previously
described by Kitagawa and colleagues [24] in a patient with a distal aortopulmonary septal defect.

As already recognized by other teams, the operative risk of a such complex operation may be similar to those found in patients with interrupted aortic arch and ventricular septal defect. Mid-term hemodynamic and clinical results seem to be very satisfactory [20]. During 4, 5 years of follow up, an early surgical correction of APW and associated lesions showed excellent survival rate, freedom from re-intervention and no evidence of persistent pulmonary hypertension post repair [25,26]. During this patient last follow up on March 2019 at NCHHK Pediatric Cardiology clinic, echo result showed no residual AP Window, minimal pressure gradient of 18 mmHg across the aortic reconstruction with good LV function with EF of 70%, good RV function, intact inter atrial & ventricular septum with TR Mild and TVG 20 mmHg, no residual pulmonary hypertension was detected (Fig. 6).

We believe strongly that the overwhelming majority of neonates with this anomaly are best served by a simple one-stage procedure through a median sternotomy involving direct aortic arch anastomosis and patch closure of the AP window. We hope that the long term outcome can be obtained well, as well as the process of growth and development of the patients at school age will be excellent.

4. Conclusion

One-stage correction of Berry syndrome in neonate is a highly risky and complex operation, but it can be safely performed in experienced centres. Echocardiography can provide adequate diagnostic information. Early surgical treatment is a therapeutic option, These patients have a good prognosis and can achieve satisfactory early and late outcomes due to advances in anaesthesia, cardiopulmonary bypass, and perioperative management. In the published literature on berry syndrome there is a very high risk of death. However in our case, even though we faced all these risks, we did not experience premature or late death. Long-term results after surgical correction are excellent regardless of age or pulmonary vascular resistance.

Declaration of competing interest

The authors declare that there is no conflict of interest regarding publication of this paper.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.102200.

Ethical approval

The informed consent form was declared from patient’s mother Mrs. Andini Kusuma (on behalf the patient), that patient data or samples will be used for educational or research purposes. Our institutional review board also do not provide an ethical approval in the form of case report.

Source of funding

No sponsorship for this case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Yopie Afriandi Habibie conceived the study, write the manuscript draft and final draft.

Pribadi Wiranda Busro, Poppy S Roebiono, Dicky Fakhri revision the final draft.

All authors played a role in conducting surgery, pre operative diagnosis with echocardiography follow up and patient care.

Registration of research studies

1. Name of the registry: not applicable
2. Unique identifying number or registration ID: not applicable
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): not applicable

Guarantor

Yopie Afriandi Habibie is the sole guarantor of this submitted article.

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Fig. 6. Long Axis Short axis view revealed no residual AP Window from 2D echocardiography and colour Doppler. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)
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