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Case Report

An operating image of an extreme case of hypoplastic left heart syndrome for instructional purposes.

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Abstract

Although Hypoplastic Left Heart Syndrome (HLHS) is a rare condition, it is nonetheless prevalent in the Middle East and East Asia. The disease is characterised by a wide fenotype range, ranging from mild mitral and aortic stenosis to severe aortic and mitral atresia. We describe a clinical example with fenotype aortic and mitral atresia, as well as an operative imaging of the disease's severe form. A 2-day-old newborn weighing 2.97 kilogrammes was admitted to our centre with a prenatal diagnosis of Hypoplastic Left Heart Syndrome.

Keywords

Hypoplastic left heart syndrome Aortic stenosis Ventricular function

Introduction

A physiological condition sonogram, like represented in literature, showed arterial blood vessel and mitral abnormality footage. On the echo continual at birth, a awfully giant arteria pulmonalis was pictured, with a awfully little aorta. The personal organiser was terribly giant and filling the aorta and therefore the aorta, right atrioventricular valve was gently meager, right bodily cavity operate sensible while not RVOTO. traditional coronary pattern. No alternative abnormal findings were detected. Born in a very protecting and specialised atmosphere at thirty six w.o., the baby was haemodynamically stable straightaway at birth and was maintaining a comparatively sensible systemic-pulmonary setting balance with eighty three of saturation. once eight hrs aged, AN infusion of alpoprostadil was started on, following severe desaturation and symptom requiring cloth, imperative introduction, mechanical ventilation and dysfunction in presence of personal organiser narrowing. The baby has been stable till the fourth day aged while not inotropes.

On the fourth day once birth, he underwent a Norwood repair by using the thus known as "Sano technique" with a five metric linear unit. recreational vehicle to PA passage, used as shunt. The classical "Sano" technique was used therein case with a distal anastomoses on the RPA, and therefore the chest was left open for thirty six hrs. post-procedure by going alittle liga-clip on the Sano shunt so as to scale back the pulmonic overflow, in sight of the pulmonic tube-shaped structure resistance falling. oajpinternational.info Patient remained stable with some milrinone drug on board and every one the very important parameters so as, compatible with the new univentricular physiological scenario. The post-operative course was placid and therefore the patient was discharged on the eighth post-operative day in fitness and stable haemodynamical standing.

Case Report

We gift the operative image taken in theatre of the case represented, once the os and therefore the serosa are opened and therefore the thymus glands entirely resected. the image show a large arteria pulmonalis compared to a filamentlike aorta surprising as compared to similar cases. The initial sensible pulmonary/systemic haemodynamical balance, hold at birth, will be explained excellent by the personal organiser filling AN enlarged arteria pulmonalis till the personal organiser narrowing once some hours once birth. The fenotype of HLHS showed within the image, represents the a lot of extreme one associated to endocardial fibro-lelastosis on echo (Figure 1). we expect that image would be helpful to students and surgeons in coaching as instructional image so as to require into consideration the surgical state of affairs long-faced on live [3-5].

Discussion

This fenotype must be thought of mutually as associated to the best peri-operative mortality and short survival on the long distance , even once the third palliative stage procedure enclosed into the standard treatment pathway.
The second surgical step, planned between 3-6 months aged, to treat AN HLHS could be a metal caval-pulmonary affiliation conjointly known as " John Glenn procedure" so as to permit {a better|a far better|a much better|a higher|a stronger|a a lot of robust|an improved} growth of the pulmonic blood vessel tree that don't seem to be well supported more by the tiny central Sano shunt applied at birth , and continue on the univentricular haemodynamical pathway, which is able to divert the blood come back on to the lungs , bypassing the recreational vehicle that is especially acting as general blood vessel pump chamber connected to the arterial blood vessel by the Damus-Stansel-Key procedure [6,7]. The completion of the univentricular pathway is pictured by the whole cavo-pulmonary affiliation according nowdays to the extra-cardiac passage by using the thus known as "Fontan procedure" between 2-4 years aged.

That final step are prosperous only a minimum of ten physiological principles are revered [8].
In some occasions, given by a not extreme fenotype of the malady, like mitral and stricture of low grade, it's doable to realize a biventricular repair, planned ahead once some initial palliative procedures. the possibility of a decent result's calculated preoperatively by the Rhode score on echo, that takes into thought totally different parameters, as well as the 55 kind and its volume in step with the anatomy, and therefore the presence of serosa fibro-elastosis malady associated to [9].
The low Rhodes score calculated on echo during this specific case, didn't provide any indication to a biventricular repair.

The severe uni-ventricular kind expressed even by a filamentlike arterial

blood vessel , during this case , sadly carries a poor prognosis on the medium term conjointly thanks to terribly high risk of re-coartation of arterial blood vessel requiring many surgical or interventional procedures , on every occasion tight and at high mortality risk [7-11].
In HLHS mortality is in any case terribly high on the future, even once a prosperous third stage palliation procedure [12-15].

The 55 itself and every one the elements of the 55 square measure severely underdeveloped here [12]. The presence of a solitary recreational vehicle, usually associated to a angulate insufficiency doesn't guarantee, on the future, a correct survival [13]. The presence of a a lot of or less recreational vehicle development , ability and anatomy continues to be debatable with regard to the future survival however looks currently quite clear that the recreational vehicle operate and anatomy square measure 2 of things of overriding importance for the future survival up to the youth (Figure 2). The physiopatology of the one heart has been all right studied over the last years, and positively that has contributed to a stronger post-operative and future survival.

Even considering that, the $64000 final ANd definitive conclusion within the treatment of an HLHS is pictured by the center transplant [15]. Also this procedure is charged of high peri-operative mortality, following the complicated and sick multi-organ failure of patients accepted into the center roster. That doesn't facilitate the selection of these candidates within the roster, once AN heart is out there for transplant, if we tend to contemplate the shortness of organs like we tend to face nowdays. A dialogue on the transplantability of the Fontan patients is for that reason presently on going. The psychological impact on the adult patients survived to the future Fontan procedure however going towards the darkness concerning their future wants conjointly to be taken into thought.

Conclusion

It is still debatable whether or not or to not intervene in any respect on babies born with HLHS or it'd be higher interrupt the gestation in Countries a minimum of wherever that's wrongfully allowed.

References

1. Bravo-Valenzuela NJ, Peixoto AB, Araujo Júnior E, et al. Prenatal diagnosis of congenital heart disease: A review of current knowledge. Indian Heart J. 2018;70:150-164.

2. Crucean A, Alqahtani A, Barron DJ, et al. Re-evaluation of hypoplastic left heart syndrome from a developmental and morphological perspective. Orphanet J Rare Dis. 2017;12:138.

3. Newland DP, Poh CL, Zannino D, et al. The impact of morphological characteristics on late outcomes in patients born with hypoplastic left heart syndrome. Eur J Cardiothorac Surg. 2019;56:557-563.

4. Plymale JM, Frommelt PC, Nugent M, et al. The Infant with Aortic Arch Hypoplasia and Small Left Heart Structures: Echocardiographic Indices of Mitral and Aortic Hypoplasia Predicting Successful Biventricular Repair. Pediatr Cardiol. 2017;38:1296-1304.

5. Abarbanell G, Border WL, Schlosser B, et al. Preoperative echocardiographic measures in interrupted aortic arch: Which ones best predict surgical approach and outcome? Congenit Heart Dis. 2018;13:476-482.

6. Linda Edwards, Kevin P Morris, Ameen Siddiqui, et al. Norwood procedure for hypoplastic left heart syndrome: BT shunt or RV-PA conduit? Arch. Disease Child Fetal Neonatal. 2007;92:210–214.

7. Sakurai T, Rogers V, Stickley J, et al. Single-center experience of arch reconstruction in the setting of Norwood operation. Ann. Thorac Surg. 2012;94:1534-9.

8. Hosein RB, Clarke AJ, McGuirk SP, et al. Factors influencing early and late outcome following the Fontan procedure in the current era. The 'Two Commandments'? Eur J Cardiothorac Surg. 2007;31:344-52.

9. Tani LY, Minich LL, Pagotto LT, et al. Left heart hypoplasia and neonatal aortic arch obstruction: is the Rhodes left ventricular adequacy score applicable? J Thorac Cardiovasc Surg. 1999;118:81-6.

10. Mart CR, Eckhauser AW. Development of an echocardiographic scoring system to predict biventricular repair in neonatal hypoplastic left heart complex. Pediatr Cardiol. 2014;35:1456-66.

11. Szypulski A, Rai V, Sacharczuk J, et al. Risk factors for recoarctation of aorta after Norwood procedure in patients with hypoplastic left heart syndrome. Folia Med Cracov. 2018;58:11- 21.

12. Bjurbom M, Iyengar AJ, Moenkemeyer F, et al. Evolution of Left Ventricular Size in Late Survivors of Surgery for Hypoplastic Left Heart Syndrome. Ann Thorac Surg. 2017;104:926-931.

13. Petko C, Möller P, Hoffmann U, et al. Comprehensive evaluation of right ventricular function in children with different anatomical subtypes of hypoplastic left heart syndrome after Fontan surgery. Int J Cardiol. 2011;150:45-9.

14. Walsh MA, McCrindle BW, Dipchand A, et al. Left ventricular morphology influences mortality after the Norwood operation. Heart. 2009;95:1238-44.

15. Roeleveld PP, Axelrod DM, Klugman D, . Hypoplastic left heart syndrome: from fetus to Fontan. Cardiol Young. 2018;28:1275-1288.