An Unusual Airway complication in a patient with Pulmonary Atresia

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Background

Pulmonary atresia with ventricular septal defect (PA/VSD) is an extremely rare congenital cardiac abnormality. Early pulmonary atresia at the time of truncoconal partitioning gives rise to VSD[1]. Treatment is dictated largely by anatomical variance. The right and left pulmonary arteries may or may not communicate, with an overriding aorta that delivers blood to both the pulmonary and systemic circulation. Patent ductus arteriosus is vital to early survival of these patients. The lungs are typically supplied by major collateral arteries arising from the abdominal/thoracic aorta and ductus arteriosus may remain patent.

Typical presentation is central cyanosis or bluish discoloration of the face at birth, although evidence can be seen at 18-22 weeks gestation on fetal ultrasound. Etiology is not fully understood but factors contributing to disease include family history, teratogenic drug usage of mother, smoking, diabetes and late pregnancy. It is decreasing in prevalence disproportionally to other congenital heart disease.

Treatment is typically either staged, or total early surgical correction. Staged correction involves anastomosis of the pulmonary arteries and aorta, before reconnection of the right ventricle to the pulmonary arteries. Finally, the Ventricular defect is repaired at 1-3 years.

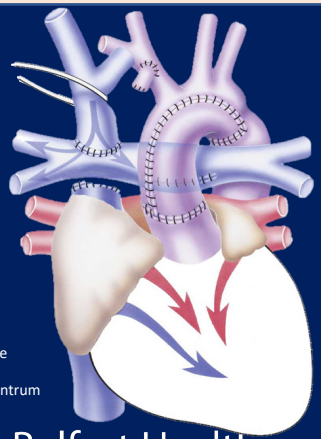
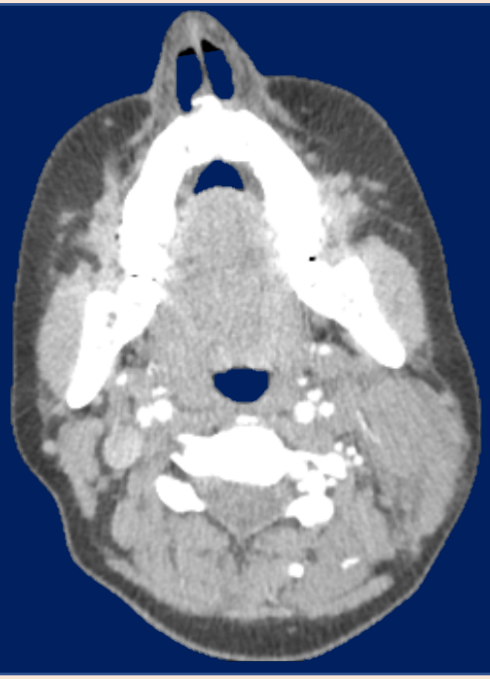


Fig 1. Bidirektionale Glenn- Operation Deutsches Herrzzentrum Berlin

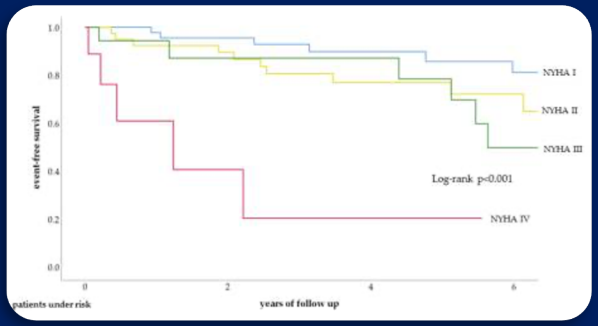


Case

A 27-year-old patient with PA/VSD presented with sudden onset right sided paraesthesia and ataxia on a background of multiple previously occluded cardiac stents and Pulmonary Emboli, for which they were receiving lifelong Warfarin. Initial Angiography revealed occlusion of the M1 and M2 branches of the Left Middle Cerebral Artery. It was decided the patient would be treated with clot retrieval. 19 hours post-procedure, the patient developed a rapidly growing, firm 7x3 cm lesion over the left anterior sternocleidomastoid area of the neck with no cutaneous change on examination. Urgent Aortic arch angiography revealed intra-sternocleidomastoid haemorrhage with supply from an unidentified proximal artery of the external carotid. Concern was raised over internal compression of the patient's oropharynx and ENT were contacted to assess the patient's airway. Flexible Nasal endoscopy revealed no internal compression of the laryngeal cavity. Warfarinisation was reversed with Vitamin K and Octaplex was administered. The patient was discharged 10 days later with improving haematoma and only mild changes on follow up CT brain.

Discussion

As a result of advancing surgical interventions since 1950, the population of patients reaching 40 with PA/VSD is increasing year on year. Incidence is also decreasing as greater understanding of teratogenic drugs during pregnancy is leading to a decrease in prevalence[2]. We have a greater understanding of comorbidities in patients approaching older ages, alongside more effective treatments e.g improved heart failure medications and implantable electrophysiology devices[3].



Purpose

To promote the importance of vigilant monitoring of patients post-procedurally with little anatomical reserve and to educate about the aging population of patients with cyanotic heart disease.

References

1. Kutsche, L. Van Mierop, L. 1983.Pulmonary atresia with and without ventricular septal defect: a different aetiology and pathogenesis for the atresia in the 2 types? *American Journal of Cardiology* ; 15;51(6):932-5.
2. Sana MK, Ahmed Z. Pulmonary Atresia With Ventricular Septal Defect. [Updated 2021 Sep 28]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK562277/
3. Hock J, Schwall L, Pujol C, et al. Tetralogy of Fallot or Pulmonary Atresia with Ventricular Septal Defect after the Age of 40 Years: A Single Center Study. *J Clin Med*. 2020;9(5):1533. Published 2020 May 19. doi:10.3390/jcm9051533 also Fig 2.

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