Symptomatic neonatal tetralogy of Fallot: repair or shunt ?

Definition

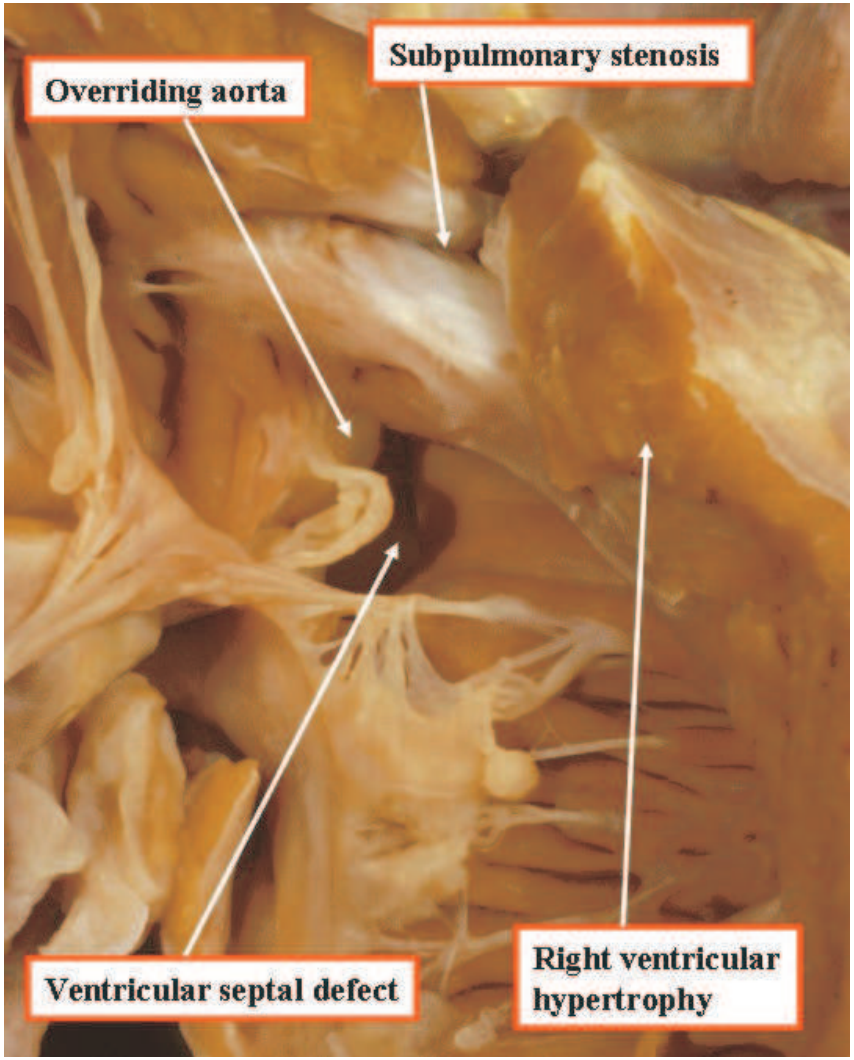
- Lesions occurs in 3 of every 10,000 live births, and accounts for 7–10% of all congenital cardiac malformations.

❖ Anatomic Features

1) RVOT obstruction (infundibular stenosis)
2) VSD
3) Aorta dextroposition, overrides VSD
4) RV hypertrophy

The VSD and infundibular stenosis determine the pathophysiologic features

Definition


Overriding aorta
Subpulmonary stenosis
Ventricular septal defect
Right ventricular hypertrophy

Controversy

- Some centres will perform complete repairs in all neonates. Others will palliate symptomatic neonates, and perform a complete repair in all patients at the age of 4 to 6 months.

Complete neonatal repair(adv)

- Prompt relief of the volume and pressure overload on the right ventricle,
- Minimises cyanosis, decreases parental anxiety, and eliminates the theoretical risk of stenosis occurring in a pulmonary artery due to a palliative procedure

- Frederique Bailliard, Robert H Anderson. ReviewTetralogy of Fallot. *Orphanet Journal of Rare Diseases 2009, 4:2*

Complete neonatal repair(adv)

- Early repair, providing adequate pulmonary blood flow, with homogeneous distribution to both lungs, allows normal growth and development of the pulmonary arteries.
- This is an advantage considering the potential negative effects on the pulmonary arteries of a systemic-to-pulmonary shunt.

Complete neonatal repair(adv)

- Patients who undergo a successful complete repair during the neonatal period will be unlikely to require more than one intervention in the first year of life, but are not free from reintervention.

Complete neonatal repair(disadv)

- Shunting or primary repair of neonates with symptomatic TOF provides equivalent mortality and results.
- The primary repair patients more frequently had a transannular patch and a tendency to more frequent delayed sternal closure .

Complete neonatal repair(disadv)

- Shunted patients had fewer transannular patch repairs despite having more emergent initial operations.
- Compared with the primary repair group, shunted patients had decreased intensive care unit and hospital stays for the first hospitalization, which were neutralized when the second operation (repair) values were added.

- Kanter KR, Kogon BE, Kirshbom PM, Carlock PR. Symptomatic neonatal tetralogy of Fallot: repair or shunt? Ann Thorac Surg. 2010 Mar;89(3):858-63.

Complete neonatal repair(disadv)

- Neonatal complete repairs include exposure of the neonatal brain to cardiopulmonary bypass, and the increased need to place a patch across the ventriculo-pulmonary junction when compared to older age at repair

- Hirsch J, Mosca R, Bove E: Complete repair of tetralogy of Fallot in the neonate. *Annals of Surgery* 2000, 232(4):508-514. 25.
- Stewart RD, Backer CL, Young L, Mavroudis C: Tetralogy of Fallot: Results of a pulmonary valve-sparing strategy. *Annals of Thoracic Surgery* 2005, 80:1431-1439

Complete neonatal repair(disadv)

- Transannular patches, create a state of chronic pulmonary regurgitation, which increases morbidity in young adults, producing exercise intolerance and ventricular arrhythmias.If left untreated, this increases the risk of sudden death

- Nollert G, Fischlein T, Bouterwek S, Böhmer C, Klinner W, Reichart B: Long-term survival in patients with repair of TOF: 36-year follow-up of 490 survivors of the first year after surgical repair. *The Journal of the American College of Cardiology* 1997, 30:1374-1383.
- Helbing WA, Roest AA, Niezen RA, Vliegen HW, Hazekamp MG, Ottenkamp J, de Roos A, Wall EE van der: ECG predictors of ventricular arrhythmias and biventricular size and wall mass in tetralogy of Fallot with pulmonary regurgitation. *Heart* 2002, 88:515-520.
- Gatzoulis MA, Till JA, Somerville J, Redington AN: Mechanoelectrical interaction in tetralogy of Fallot, QRS prolongation relates to right ventricular size and predicts malignant ventricular arrhythmias and sudden death. *Circulation* 1995, 92(2):231-237.

Complete neonatal repair(disadv)

- The effect of cardiopulmonary bypass on the neonatal brain, and the associated risk of longer stay in hospital and the intensive care unit, is not trivial.
- Studies of neurodevelopmental outcomes of neonates undergoing cardiopulmonary bypass compared to older children have shown lower intelligence quotients in patients exposed to bypass as neonates
- Miller G, Tesman JR, Ramer JC, Baylen BG, Myers JL: Outcome after open-heart surgery in infants and children. *Journal of Child Neurology* 1996, 11(1):49-53

Complete neonatal repair(disadv)

- Longer periods of bypass, and longer stays in the intensive care unit, have been associated with an increased risk for neurological events and abnormal neurological findings on followup

- Limperopoulos C, Majnemer A, Shevell MI, Rohlicek C, Rosenblatt B, Tchervenkov C, Darwish HZ: Predictors of developmental disabilities after open heart surgery in young children with congenital heart defects. *The Journal of Pediatrics* 2002, 141(1):52-58.
- Fallon P, Aparício JM, Elliott MJ, Kirkham FJ: Incidence of neurological complications of surgery for congenital heart disease. *Archives of Disease in Childhood* 1995, 72:418-422

Complete neonatal repair(disadv)

- While some studies have not found cyanosis itself to be responsible for cognitive problems in children with congenitally malformed hearts

- Wright M, Nolan T: Impact of cyanotic heart disease on school performance. *Archives of Disease in Children* 1994, 71:64-70.

Complete neonatal repair(disadv)

- Others have implicated chronic cyanosis as a factor contributing to impaired motor skills, decreased academic achievement, and worsened behavioural outcomes

- Stieh J, Kramer HH, Harding P, Fischer G: Gross and fine motor development is impaired in children with cyanotic congenital heart disease. *Neuropediatrics* 1999, 30(2):77-82.
- Bass JL, Corwin M, Gozal D, Moore C, Nishida H, Parker S, Schonwald A, Wilker RE, Stehle S, Kinane TB: The effect of chronic or intermittent hypoxia on cognition in childhood: a review of the evidence. *Pediatrics* 2004, 114:805-816.

Difficulty

- In the absence of randomised control trials, assessing the risk and benefits of the two surgical strategies has been notoriously difficult

Summary

- Patients with cyanotic tetralogy of Fallot will either undergo neonatal complete repair or neonatal palliation with an aortopulmonary shunt followed by a complete repair at four to six months of age.
- Peri-operative mortality rates for either surgical approach is less than 3% and since neither strategy has shown superior results, surgical management remains dependent on the protocols preferred by the individual centres.

Summary

- In any case the potential advantages of a primary early repair should be weighted against the experience and expertise of the individual centre and/or surgical team in dealing with tetralogy of Fallot and with neonates and infants

Questions

- Questions that remain regarding the management of tetralogy of Fallot pertain to the preference of surgical timing,as discussed above, and to the residual lesions of the disease.
- The management of young adults with pulmonary regurgitation or residual pulmonary stenosis is complex

Answers

- Recent advances in interventional cardiac catheterisation now provide safe options for treatment other than cardiac surgery

- Lurz P, Coats L, Khambadkone S, Nordmeyer J, Boudjemline Y, Schievano S, Muthurangu V, Lee TY, Parenzan G, Derrick G, Cullen S, Walker F, Tsang V, Deanfield J, Taylor AM, Bonhoeffer P: Percutaneous pulmonary valve implantation. Impact of evolving technology and learning curve on clinical outcome. *Circulation* 2008, 117:1964-1972.
- 45. Frigiola A, Tsang V, Nordmeyer J, Lurz P, van Doorn C, Taylor AM, Bonhoeffer P, de Leval M: Current approaches to pulmonary regurgitation. *European Journal of Cardiothoracic Surgery* 2008, 34(3):576-580.