Pulmonary regurgitation after repair of Tetralogy of Fallot - Questions

1 Question 1
After pulmonary valve replacement, the following often occur

- Right ventricular ejection fraction increases
- QRS duration decreases
- Exercise tolerance increases
- Right ventricular end-diastolic volume is unchanged
- Ventricular arrhythmias are abolished

1.1 Answer 1

The response to pulmonary valve replacement is very variable, and to a considerable extent depends on whether right ventricular dilatation is reversible and how long after valve replacement the assessment is made.

- True. Right ventricular ejection fraction corrected for pulmonary and tricuspid regurgitation often increases following pulmonary valve replacement (1, 2). Right ventricular ejection fraction is more frequently reported uncorrected for regurgitant flows, and typically does not increase following valve replacement. However, Therrien et al. (3) found that uncorrected right ventricular ejection fraction increased in individual patients following pulmonary valve replacement even though there was no change in the group as a whole.

- True. Shortening of the QRS may occur, but once the new valve becomes leaky prolongation may recur.

- True.

- True.

- False. In terms of arrhythmia reduction, the current best results seem to be obtained when pulmonary valve replacement is combined with intra-operative cryoablation. However recurrence still occurs in approximately 10-15%.

2 Question 2
Surgery for tetralogy of Fallot

- Complete repair is best performed in the neonatal period independent of symptoms
• A transannular patch can be avoided in the vast majority of patients

• The surgeon encounters little variability in the right ventricular outflow tract or pulmonary artery anatomy

• Patients who receive a pulmonary valvotomy, but do not require a transannular patch, do not develop clinically significant pulmonary regurgitation.

• A mechanical valve is the valve of choice for pulmonary valve replacement

2.1 Answer 2

Despite being one of the oldest congenital heart operations, many aspects of the surgical management of Tetralogy of Fallot remain contentious.

• False. The overall trend is to perform elective repair in the first year of life e.g. at approximately six months of age. These patients have acceptable transannular patching rates and the majority have very short intensive care unit stays. Given the heterogeneity in Tetralogy anatomy good results may be obtained in individual patients even in early infancy. The real debate relates to the management of the severest end of the spectrum, the cyanotic neonatal Fallot, and the optimal strategy is unclear at present. However institutions that follow a blanket policy of neonatal complete repair for cyanotic and non-cyanotic patients usually report high transannular patching rates and long intensive care unit stays e.g 10-14 days.

• False, at present. The typical transannular patching rate in unselected cohorts of Tetralogy patients is approximately 50%. In small series this has been reported to be as low as approximately 30%. Considerable effort is being expended to decrease the transannular patching rate, even at the expense of some residual stenosis.

• False

• False. It remains to be proven that those with valvotomies have a slower rate of progression of right ventricular dilatation than those with transannular patches. See Question 4.

• False

3 Question 3

The left ventricle in tetralogy of Fallot with pulmonary regurgitation

• The LV lateral wall is activated later than the right ventricle if RBBB is present

• LV dysfunction has little impact on exercise tolerance

• Diastolic ventricular interdependence only occurs in those hearts with a residual VSD

• Diastolic ventricular interdependence can arise from bowing of the left ventricular septum into the left ventricle.

• May be assisted by resynchronisation therapy
3.1 Answer 3

- False
- False
- False
- True
- True, possibly. This is an active area of research and the exact indications and magnitude of clinical benefit remain to be determined.

4 Question 4

Clinically significant pulmonary regurgitation may occur in

- Valvar pulmonary stenosis after balloon valvuloplasty
- Patients who have undergone a Rastelli operation
- Patients who have had already had a pulmonary valve replacement
- Repaired common arterial trunk
- Congenital isolated pulmonary regurgitation

4.1 Answer 4

Any patient with a right ventricle to pulmonary artery conduit has the potential to develop clinically significant pulmonary regurgitation.

- True. Clinically significant pulmonary regurgitation can even occur in patients late after surgical pulmonary valvotomy or balloon dilatation of the pulmonary valve.
- True
- True
- True
- True

5 Question 5

Management of pulmonary regurgitation

- The mainstay of treatment is drug therapy
- In a patient with a pulmonary regurgitant fraction of 53 %, a Doppler peak gradient across the right ventricular outflow tract of 40 mmHg suggests there is very likely to be a severe stenosis
- Tricuspid regurgitation is an early sign of significant pulmonary regurgitation
- Regular Holter monitoring is useful
- There is little benefit from relieving branch pulmonary artery stenoses
5.1 Answer 5

- False
- False. Right ventricles with a large stroke volume frequently have moderate Doppler derived gradients across the outflow tract without anatomical narrowing.
- False. In general, tricuspid regurgitation is a late sign of pulmonary regurgitation.
- False. Holter monitoring is useful in the investigation of symptoms.
- False. Relieving branch pulmonary artery stenoses can decrease pulmonary regurgitation.

6 Question 6

- In most tetralogy patients with pulmonary regurgitation, the majority of the regurgitant volume comes from the left pulmonary artery
- In the assessment of pulmonary regurgitation by magnetic resonance imaging, pulmonary regurgitant fraction is a much more useful measurement for clinical decision making than right ventricular end-diastolic volume
- The best candidates for the Bonhoeffer pulmonary valve are those patients who received a transannular patch in their native right ventricular outflow tract
- The Bonhoeffer pulmonary valve cannot be deployed in a branch pulmonary artery
- In a very cyanosed neonate with Ebstein’s anomaly with severe tricuspid regurgitation and no right ventricular outflow tract obstruction, maintaining ductal patency is critical to survival

6.1 Answer 6

- True
- False. The regurgitant fraction is a ratio, and is sensitive to changes in both antegrade and retrograde flow volumes. It is easy to envisage a situation with static valve anatomy, where the regurgitation fraction decreases with time as the right-ventricle progressively dilates and the right ventricular distolic pressure profile alters. Although it is more difficult to measure, right ventricular dilatation as assessed by the right ventricular end-diastolic volume is the more important quantity as it reflects the important consequence of pulmonary regurgitation
- False. In the current Bonhoeffer system, the best implantation sites are conduits or homografts of 22 mm diameter or less.
- False. A small number of patients have had valves successfully delivered into a branch pulmonary artery. The long-term outcome of this strategy and its effect on the total regurgitant volume from the contralateral pulmonary artery are unclear, at present.
• False. Ductal ligation can be dramatically beneficial, by removing the driver of the circular shunt.

References

