Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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CASE SUMMARY

The 28-year-old Caucasian male, active, sport teacher, with no remarkable family history, who underwent the total correction of tetralogy of Fallot in his early childhood. He was admitted to our Center of Rare Cardiovascular Diseases due to episode of the syncope with loss of consciousness during the skiing. In the medical review he got over surgical correction of residual ventricular septal defect in 1992, then percutaneous balloon angioplastic of constricted left branch of pulmonary artery in 2000 year. The patient was under the observation of Pediatric until he reached 18 years old . 12–lead ECG showed sinus rhythm, 80/min, right His bundle branch block. The transthoracic echocardiography showed normal size of the left heart chambers, with preserved systolic function, enlarged size of right ventricle and atrium with moderate pulmonary and tricuspid regurgitation. The MRI of the heart was planned which revealed impaired RV contractility, moderate pulmonary valve stenosis with accelerated blood flow and right pulmonary artery dilatation, gradient in ostium about 20 mmHg. There were no visible areas of fibrosing of RV muscle. Patient had also Holter-ECG which did not revealed any significant ventricular or supraventricular arrhythmias.

DISCUSSION

The occurrence of sudden death has been recognized as a late complication of surgical correction of tetralogy of Fallot. The incidence of sudden death appears to be between 1.5 and 4.5 deaths per 1000 patient years, most commonly occurring 4 years or more after repair. In spite of the number of techniques available, no single risk factor has yet been identified to predict VT and sudden death convincingly in corrected tetralogy of Fallot. Follow-up of subjects with tetralogy of Fallot for evaluation of susceptibility to ventricular arrhythmia requires consideration of clinical history, details of operative procedures undertaken, post-procedural catheterization data, ECG data and ongoing non-invasive haemodynamic assessment. Once an at-risk patient is identified, the optimum management strategy remains unclear, with the probable exception that anti-arrhythmic medication in the long-term may present an inadequate solution. However, the development of successful catheter ablation, the availability of automatic implantable cardio-defibrillators, and the potential reduction in arrhythmia propensity following pulmonary valve replacement, mean that therapeutic strategies are available [1,2,3,4].

Risk is affected by the type of repair performed: use of a right ventriculotomy is associated with the highest rate of ventricular dysrhythmia, and the presence of a ventricular septal patch is a second potential source of reentrant arrhythmia. Fibrous–fatty substitution occurs at the site of surgical scarring, and may provide a substrate for abnormalities of depolarization and repolarization. The quality of surgical correction affects long-term susceptibility to sudden death, with residual outflow obstruction and pulmonary regurgitation of particular importance. Post-operative cardiac catheterization identifies patients at risk of ventricular arrhythmias (RVSP>60mmHg) and patients at risk of sudden death (residual RV to PA outflow tract gradient >40mmHg). Post-operative complications increase the frequency of late events. Transient complete heart block in the immediate post-operative period, despite return to sinus rhythm before discharge, is associated with a five-fold increase in late sudden death-presumably because of injury to the conduction system [1,2].

EXPERT'S OPINION

If there is no obvious arrhythmias diagnosed antiarrhythmic intervention and normal cardiac contractility, i.e. ICD implantation seems unnecessary. Optimal pharmacological therapy should be recommended now.

CONCLUSION

Taking into consideration his complete clinic picture - symptoms – one episode of syncope with no evidence of arrhythmia, generally good tolerance of physical activity he was decided to be treated conservatively and to be under continuous observations. It was also concluded that replacement of stenotic pulmonary valve should be considered as far as symptoms occurred. The ICD implantation as a primary sudden cardiac death prevention is not necessary at least at this time. Close observation with regular exercise tolerance evaluation - repeated Cardiac echo, Holter ECG is needed.

REFERENCES

1. Steeds RP, Oakley D. Predicting late sudden death from ventricular arrhythmia in adults following surgical repair of tetralogy of Fallot. QJM: An International Journal of Medicine 2004; 97: 7-13. doi: 10.1093/qjmed/hch004

2. Gatzoulis MA, Balaji S, Webber SA**,** et al. Risk factors for arrhythmia and sudden cardiac death late after repair of Tetralogy of Fallot: a multicentre study. Lancet 2000; 356: 975 - 81.

3. Helbing WA, Roest A, Niezen RA, et al. ECG predictors of ventricular arrhythmias and biventricular size and wall mass in tetralogy of Fallot with pulmonary regurgitation. Heart 2002; 88: 515 - 519.

4. Le Gloan L., Khairy P. Management of arrhythmias in patients with tetralogy of Fallot. Current Opinion in Cardiology 2011; 26: 60–65.

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