

Medical Expertise

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

EXPERT: Dr Bartosz Sobień, cardiologist

Affiliation: *Department of Cardiac and Vascular Diseases, John Paul II Hospital, Krakow, Poland*

CASE SUMMARY

A patient with Tetralogy of Fallot, after a multistep treatment of cardiac i.e. bilateral fusion Blalock-Taussig shunt method (1988, 1989), the total correction of Fallot (1993), after homograft implantation of pulmonary and tricuspid valvuloplasty way De Vega due to severe valvular regurgitation: pulmonary and tricuspid (2007), with ventricular arrhythmias, paroxysmal atrial flutter, atrioventricular distraction.

DISCUSSION

Anatomy of the conduction system in patients with ToF is usually correct. Sinus node lies in the wall of the right atrium near the junction of the superior vena cava and the atrioventricular node in the posterior triangle of Koch. Bunch branch extends through atrioventricular septum and in the level of the crest of the muscular portion of the septum is divided into two legs. Right leg, initially running intramuscularly appears subendocardial near posterior lower edge formed as a result of abnormal development of the cone chambers, ventricular septal defect type malalignment. It is particularly vulnerable to surgical trauma. Performed during the correction of the total right-sided defects ventriculotomy in most cases results in a right bundle branch block. Depending on where the damage may be the central or peripheral block formed as a result of damage to the Purkinje fibers; may also be the result of both these components. The present RBBB usually has the characteristic morphology of: short and narrow first arm R1, and high and wide arm R2. In the current studies it has been demonstrated that increasing exercise early treatments correction of the total ToF at increasingly younger patients does not increase the frequency of the occurrence of significant conduction disturbances. Atrioventricular block third degree may occur in the short postoperative period or many years after surgery. It has been proved that the block late often occur in patients who have seen early, transient postoperative total block. The consequence of

the presence of atrioventricular block third degree is a need for constant electrical stimulation. Adults with congenital heart defects are a specific group of patients - a lifelong addicted to constant electrical stimulation.

EXPERT'S OPINION

Disorders of atrioventricular conduction and / or sick sinus syndrome do not occur in the adult population of patients after total correction of ToF, but often they are a significant clinical problem that requires close observation, especially that by degenerative aging phenomenon will intensify. The consequence of the presence of atrioventricular block third degree is a need for constant electrical stimulation.

CONCLUSION

In the case of persistent conduction disorders - ventricular pacemaker implantation is recommended .

REFERENCES

1. Trojnarowska O, Łanocha M, Pyda M, et al. Disturbances of conduction and trigger in adult patients in long-term follow up after total correction of tetralogy of Fallot. *Folia Cardiol.* 2006, tom 13, nr 2, 147–153.