Resynchronization therapy for failing sub-pulmonary right ventricle in congenital heart disease

Jan Janoušek
Children's Heart Center, 2nd Faculty of Medicine, Charles University in Prague and Motol University Hospital, Prague, Czech Republic

Pulmonary right ventricular (RV) dysfunction is present in a number of congenital heart lesions after repair surgery. It is frequently associated with right bundle branch block (RBBB) as a source of electromechanical dyssynchrony which may contribute to RV failure. RV cardiac resynchronization therapy (RV-CRT) may be used to improve RV function, electromechanical synchrony and contraction efficiency. It is effective in the immediate postoperative period as a temporary treatment option. Limited evidence also indicates the effectiveness of permanent RV-CRT. Further studies are needed to better define its role in long-term RV failure management.

Klíčová slova: congenital heart disease, right ventricular dysfunction, right ventricular dyssynchrony, bundle branch block, cardiac resynchronization therapy.

Resynchronizační léčba selhávající subpulmonální pravé komory u vrozené srdeční vady

Dysfunkce subpulmonální pravé komory (PK) je přítomna u celé řady vrozených srdečních vad po chirurgické korekci. Je často spojena s blokádou pravého Tawarova raménka (RBBB) jako zdrojem elektromechanické dyssynchronie, která se může podílet na selhání PK. Srdeční resynchronizační léčba pravé komory (RV-CRT) se používá ke zlepšení funkce PK, elektromechanické synchronie a efektivity kontrakce. Je účinná v období bezprostředně po operaci jako dočasná možnost léčby. Omezené množství důkazů rovněž svědčí o účinnosti permanentní RV-CRT. Abychom lépe definovali její roli v léčbě dlouhodobého selhávání PK, bude zapotřebí dalších studií.

Key words: vrozená srdeční vada, dysfunkce pravé komory, dyssynchronie pravé komory, blokáda Tawarova raménka, srdeční resynchronizační léčba.

Introduction

Pulmonary right ventricular (RV) dysfunction and occasional failure are associated with a number of congenital heart lesions, most predominantly but not limited to postoperative tetralogy of Fallot (ToF). They are attributed to several factors including myocardial fibrosis due to preoperative hypoxemia and pressure overload, surgical scar and long-term post-repair volume overload caused by pulmonary regurgitation and frequently associated with right bundle branch block (RBBB) (1). RBBB is also the most frequent cause of electromechanical discoordination in congenital heart disease. In postoperative ToF, RV electromechanical discoordination has been associated with decreased RV ejection fraction, mechanical inefficiency and impaired exercise capacity (2, 3) and is hypothesized to play a significant role in RV dysfunction progression. A recently published study using computer modeling showed that the extent of negative influence of RV discoordination may be greater than previously thought and overweigh pulmonary regurgitation during exercise (4). Relief of RV volume overload by pulmonary re-valvulation is thought to reverse pathologic RV remodeling occurring due to pulmonary regurgitation in post-repair ToF patients. However, a decreased probability of reverse remodeling has been reported in patients with high RV end-diastolic and end-systolic volumes, low RV ejection fraction, and those with wide QRS complex (≥ 160 ms) (5). Thus, pulmonary valve replacement alone may not lead to RV performance normalization.

KORESPONDENČNÍ ADRESA AUTORA: prof. Jan Janoušek M.D., Ph.D., jan.janousek@fnmotol.cz
Children's Heart Center, University Hospital Motol
V Úvalu 84, 150 06 Prague 5

Cit. zkr: Interv Akut Kardiol 2019; 18(3, Suppl. A): 34–35

Cardiac resynchronization therapy (CRT) has so far been used to treat dyssynchrony in the failing systemic (left) ventricle. Robust clinical data demonstrate a favorable CRT effect on acute hemodynamics, reverse cardiac remodeling, functional capacity, and patient survival. Limited evidence suggests a similar CRT effect in children and patients with congenital heart disease (6). Reports on sub-pulmonary RV resynchronization (RV-CRT) are scarce. In this report, we would like to briefly review current experience with RV-CRT in patients with congenital heart disease.

Temporary RV-CRT

The data published show that acute RV-CRT through either RV or biventricular pacing improves short-term hemodynamics in both children and adults with congenital heart disease and RBBB, and has been successfully used to treat right ventricular dysfunction in the acute postoperative setting (7). Pacing is applied by temporary epicardial pacing wires placed by the surgeon on the right atrium and the late activated free wall of the right ventricle. RV resynchronization is achieved by atrial-triggered RV free wall pacing in complete fusion with spontaneous ventricular activation to produce maximal QRS duration shortening. A specific attention is paid to complete abolition of the broad S wave in standard lead I. The aim is to resynchronize the septum and RV free wall electrically as well as mechanically. A commercially available external dual-chamber pacemaker may be used. Temporary RV-CRT performed in this way carried a significant short-term improvement in hemodynamics in children early after surgery for ToF and may be a useful non-pharmacologic adjunct to the management of hemodynamically compromised patients. Resynchronization effect was maximized when pacing from the area of the latest RV activation. In another study temporary, RV-CRT carried multiple positive effects on RV mechanics, synchrony and contraction efficiency (8).

Permanent RV-CRT

The use of permanent RV-CRT to treat RV dysfunction associated with RBBB seems to be a further logical step. Unfortunately, published evidence is limited. In the first well documented report on permanent RV-CRT in a patient late after repair of ToF (9) both right and left ventricular function improved significantly within six months accompanied by improvement in the NYHA class as well as exercise capacity. Data on a small series of congenital heart disease patients undergoing RV-CRT have been presented in an abstract form confirming multiple positive mid-term resynchronization effects on right ventricular systolic function, electromechanical synchrony, and contraction efficiency (10).

Conclusion

RV-CRT appears to be a valuable method to treat both acute and chronic sub-pulmonary right ventricular dysfunction and failure associated with RBBB in patients with congenital heart disease. Further studies are needed to better define its role in long-term RV failure management.

Supported by the Agency for Medical Research, Ministry of Health of the Czech Republic [grant No 15-28029A] and by the project (Ministry of Health, Czech Republic) for conceptual development of research organization 00064203 (University Hospital Motol, Prague, Czech Republic).

REFERENCES

1. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, Gatzoulis MA, Gohlke-Baerwolf C, Kaemmerer H, Kilner P, Meijboom F, Mulder BJ, Oechslin E, Oliver JM, Serraf A, Szatmari A, Thaulow E, Vouhe PR, Walma E; Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC); Association for European Paediatric Cardiology (AEPC); ESC Committee for Practice Guidelines (CPG). ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010; 31: 2915–2957.

2. Hui W, Slorach C, Dragulescu A, Mertens L, Bijnens B, Friedberg MK. Mechanisms of right ventricular electromechanical dyssynchrony and mechanical inefficiency in children after repair of tetralogy of fallot. Circ Cardiovasc Imaging 2014; 7: 610–618.

3. Yim D, Hui W, Larios G, Dragulescu A, Grosse-Wortmann L, Bijnens B, Mertens L, Friedberg MK. Quantification of Right Ventricular Electromechanical Dyssynchrony in Relation to Right Ventricular Function and Outcomes in Children with Repaired Tetralogy of Fallot. J Am Soc Echocardiogr 2018; 31: 822–830.

4. Lumens J, Fan CS, Walmsley J, Yim D, Manlhiot C, Dragulescu A, Grosse-Wortmann L, Mertens L, Prinzen FW, Delhaas T, Friedberg MK. Relative Impact of Right Ventricular Electromechanical Dyssynchrony Versus Pulmonary Regurgitation on Right Ventricular Dysfunction and Exercise Intolerance in Patients After Repair of Tetralogy of Fallot. J Am Heart Assoc 2019; 8: e010903. doi: 10.1161/JAHA.118.010903. PubMed PMID: 30651018.

5. Geva T, Gauvreau K, Powell AJ, Cecchin F, Rhodes J, Geva J, del Nido P. Randomized trial of pulmonary valve replacement with and without right ventricular remodeling surgery. Circulation. 2010; 122(Suppl 1): S201–S208.

6. Motonaga KS, Dubin AM. Cardiac resynchronization therapy for pediatric patients with heart failure and congenital heart disease: a reappraisal of results. Circulation 2014; 129: 1879–1891.

7. Vojtovic P, Kucera F, Kubus P, Gebauer R, Matejka T, Tlaskal T, Lozek M, Kovanda J, Janousek J. Acute right ventricular resynchronization improves haemodynamics in children after surgical repair of tetralogy of Fallot. Europace 2018; 20: 323–328.

8. Janoušek J, Kovanda J, Ložek M, Tomek V, Vojtovič P, Gebauer R, Kubuš P, Krejčíř M, Lumens J, Delhaas T, Prinzen F. Pulmonary Right Ventricular Resynchronization in Congenital Heart Disease: Acute Improvement in Right Ventricular Mechanics and Contraction Efficiency. Circ Cardiovasc Imaging. 2017; 10: pii: e006424. doi: 10.1161/CIRCIMAGING.117.006424.

9. Kubuš P, Materna O, Tax P, Tomek V, Janoušek J. Successful permanent resynchronization for failing right ventricle after repair of tetralogy of Fallot. Circulation. 2014;130:e186–190.

10. Janousek J, Kovanda J, Lozek M, Tomek V, Gebauer R, Kubus P. Cardiac Resynchronization Therapy For Treatment Of Chronic Pulmonary Right Ventricular Dysfunction In Congenital Heart Disease. HeartRhythm 2018; 15(suppl): S270 (abstract).