British Congenital Cardiac Association

1 CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES: INDICATIONS FOR CRT?

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Background Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart defect. Late complications include systemic ventricular failure, systemic AV valve regurgitation, AV conduction problems and arrhythmias. Heart Rhythm Society (HRS) published expert consensus statement regarding arrhythmia management in congenital heart disease, including Cardiac Resynchronisation Therapy (CRT) indications for adult congenital heart disease (ACHD) patients. These include EF <35% with RV dilatation, NYHA II-IV, RBBB, QRS duration >150 ms in non-paced and if paced NYHA I-IV and >40% V-pacing

Method Review of notes for Class IIa indications for CRT device in ccTGA.

Results 55 patients were identified; mean age was 43.6±16 years.

36 patients (65%) had impaired systemic ventricular dysfunction on echo, mild (n=10), moderate (n=20) or severe (n=6). 31 (56%) of all patients had systemic AV valve regurgitation: mild (n=12), moderate (n=15) or severe (n=4). 12 (22%) patients have undergone systemic AV valve replacement surgery.

Rhythm was sinus (60%, n=33) or paced (n=15) on ECG. 5 (9%) were in atrial fibrillation, 4 (7%) 1st degree heart block. 18 (30%) had a simple pacemaker and 2 a CRT device.

4 (7%) fulfilled Class IIa indications for a CRT device, of which 2 had a simple pacemaker

Conclusion Only a small proportion of ccTGA patients fulfil Class IIa indications for CRT. However, complex anatomy and previous surgery may render implantation difficult. Each case needs to be assessed individually with combined electrophysiology and ACHD input with care translating guidelines into clinical practice.

2 SUITABILITY OF UNIVENTRICULAR PATIENTS FOR CARDIAC RESYNCHRONISATION THERAPY

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Introduction In 2014 the Paediatric And Congenital Electrophysiology Society (PACES) and Heart Rhythm Society (HRS) published expert consensus statement regarding arrhythmia management in congenital heart disease, including Cardiac Resynchronisation Therapy (CRT) indications for adult congenital heart disease (ACHD) patients.

These guidelines recommend CRT in univentricular hearts for those who have an impaired ventricle (EF ≤35%) with QRSd ≥150 ms, ventricular dilatation and NYHA II-IV [IIa, C].

Methods Retrospective analysis for EF, QRSd and NYHA status, from 211 patients with a Fontan circulation under specialist ACHD follow-up.

Results Univentricular functional data was available for 94% (198, 84 CMR and 114 echo). 10 (5%) patients had EF ≤35%. QRSd was available for 91% (193). 5 (3%) had QRSd≥150 ms.

2 patients were identified with both EF ≤35% and QRSd ≥150 ms, fulfilling indications for CRT. One patient was NYHA IV, tricuspid atresia with an atriopulmonary fontan and subsequent conversion to total cavopulmonary circulation (TCPC). The second patient (NYHA II) had right atrial isomerism with pulmonary atresia and an unfenestrated TCPC.

Conclusions If recent HRS CRT guidelines are applied to this large adult Fontan population there are very few candidates for CRT. Guidelines should be interpreted with caution. In virtually all cases, CRT device implantation in the adult Fontan will require a cardiac surgical approach associated with significant morbidity and mortality. The decision to implant CRT in Fontan patients is therefore complex and requires discussion in a combined ACHD electrophysiology MDT with careful consideration of patient anatomy and previous surgery

3 IMPLANTABLE LOOP RECORDERS IN ADULT CONGENITAL HEART DISEASE: A SINGLE-CENTRE EXPERIENCE

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Introduction Evaluation of rhythm dise ase can be difficult in patients with adult congenital heart disease (ACHD), especially if symptoms are infrequent or atypical, further compounded by the presence of electrocardiogram abnormalities. Implantable loop recorders (ILRs) are useful in evaluating these disorders and correlating ECG abnormalities with patient symptoms. We therefore sought to evaluate the use of ILRs in our ACHD population.

Methods A single-centre retrospective case note review of all our ACHD patients who had insertion of an ILR.

Results 119 patients (median age [37 years 29 – 48], 54 male) received an ILR, 60 complex (eg. Fontan, Ebstein, rTOF, Senning, Mustard) and 59 simple. Indications included syncope 52 (44%), pre-syncope 15 (13%), palpitations 36 (30%), prior to consideration of PFO closure 4 (3%) and other 4 (3%) and unknown 8 (7%). It took a median of 97 (52 – 200) days before a decision was made about whether symptoms correlated with a rhythm disorder or not. Outcomes include 18 (15%) significant conduction disease requiring a pacemaker, 21 (17%) patients had evidence of a new tachyarrhythmia on interrogation of the ILR, with 4 (3%) receiving an implantable cardiac defibrillator. In 41 (34%) patients the ILR was able to confirm that symptoms correlated with normal heart rhythm.

Conclusions ILRs provided a useful tool to investigate ACHD patients with symptoms suggestive of heart rhythm disorders. Identifying these patients led to timely potentially life-saving therapy.