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Matthew O'Connor, MB BChir, S. Yen Ho, PhD, Karen McCarthy, PhD, Michael Gatzoulis, MD PhD, Tom Wong, MBChB

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Short Title:
Left bundle pacing in TGA post-Mustard repair

Authors:
Matthew O’Connor MB BChir\textsuperscript{1,2}
S. Yen Ho PhD\textsuperscript{3}
Karen McCarthy PhD\textsuperscript{3}
Michael Gatzoulis MD PhD\textsuperscript{2}
Tom Wong MBChB\textsuperscript{1,2} *

\textsuperscript{1} Department of Electrophysiology, Royal Brompton Hospital, London, UK
\textsuperscript{2} Adult Congenital Heart Centre, Royal Brompton Hospital, London, UK
\textsuperscript{3} Cardiac Morphology Unit, Royal Brompton Hospital, London, UK

*Corresponding author: Tom Wong, Royal Brompton and Harefield Hospitals, Guy’s and St Thomas’ NHS Foundation Trust, Sydney Street, London, SW3 6NP twong@rbht.nhs.uk

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Introduction:

Transposition of the great arteries (d-TGA) is a congenital heart defect characterised by concordant atrioventricular and discordant ventriculoarterial connections leading to two parallel circulatory systems. There are a number of operations that have been performed over the years to surgically palliate this parallel circulation; currently the arterial switch operation is the standard of care. There are occasions where the arterial switch operation is not feasible (for example due to hostile coronary anatomy) and alternative surgical approaches such as the more historical Mustard/Senning operations must be used. These atrial switch operations were first successfully performed in 1952 and there continues to be a large population of patients with such anatomy1. In short, the Mustard operation consists of the superior vena cava (SVC) and inferior vena cava (IVC) being re-routed via surgical baffle to the vestibule of the left atrium (LA), with the subsequent re-direction of systemic venous blood to the sub-pulmonary left ventricle (LV). The remaining pulmonary venous component of the LA is re-directed rightward through the right atrium (RA) into the systemic right ventricle (RV).

A significant proportion of patients who have undergone an atrial switch operation will develop complete AV block and require pacing2. Furthermore, up to 25% will develop systemic RV failure which is precipitated or exacerbated by chronic ventricular pacing3,4. Cardiac resynchronisation therapy (CRT) has been shown to prevent or improve LV dysfunction in appropriately selected patients with normal cardiac anatomy5 and its use has extended with evidence of symptomatic benefit into the adult congenital heart disease population. Importantly the coronary sinus is inaccessible via a venous approach in 50% of
atrial switch patients as the ostium may drain into the pulmonary venous atrium post-operatively.

His-bundle pacing has gained traction as an alternative to CRT with a rapidly emerging evidence base and inclusion in major guidelines\textsuperscript{6,7}. Elevated thresholds have been reported during follow up of His-bundle leads; the aetiology of this finding is not established, but is potentially due to lead micro-dislodgement or direct injury to the His bundle. Left bundle pacing (whereby the pacing lead is tunneled through the ventricular septum to the left bundle area) has developed as a viable alternative\textsuperscript{8}. Due to the position deep in the septum the issues of both micro-dislodgement and direct His-bundle injury are addressed. It has also been employed in the ACHD population such as congenitally corrected TGA\textsuperscript{9}. However, the implant complexity increases in d-TGA, particularly as implant tools – namely the C315 sheath (Medtronic, Minneapolis, USA) – are designed to implant a lead into a septum in the usual anatomical arrangement where the ventricular septum is located posteriorly within the right ventricle instead of the anterior angulation required when implanting via a sub-pulmonary LV. We describe the technique of left bundle pacing in a patient with d-TGA and a previous atrial switch operation guided by electroanatomical mapping.

Case:

A 61-year-old man with a history of d-TGA and an atrial switch operation (Mustard) at the age of 12 who had been lost to follow up for 18 years presented to an outside centre with breathlessness, fatigue and prominent peripheral venous dilation. A 12-lead ECG demonstrated complete heart block with underlying sinus rhythm, QRS duration 142ms with a right bundloid morphology (Figure 1). Holter monitoring revealed high grade AV block with >5 second pauses during waking hours. Cardiac MRI demonstrated stenosis of both SVC and IVC baffles with mildly impaired systemic RV function (EF 56%) and that the
coronary sinus drained into the pulmonary venous atrium. The baffles were percutaneously
stented with two 39mm CP stents (NuMED Inc., Hopkinton, USA) via the right internal
jugular vein and the right femoral vein with excellent angiographic and haemodynamic result.

The patient then attended the electrophysiology laboratory two days later for
implantation of a dual chamber, left bundle area pacemaker under general anaesthesia. Due to
the angulation of the baffles, inferior access for mapping was considered unfavourable and
access was via the right basilic vein. The basilic vein measured 29mm in diameter and
accepted a 6Fr and an 8Fr sheath placed via the modified Seldinger technique under
ultrasound guidance. A fixed curve quadrapolar catheter (St Jude Medical, Minnesota, USA)
was placed in the sub-pulmonary LV apex to provide back-up pacing.

The 3D electroanatomical mapping system, EnSite Precision with the Advisor HD
grid mapping catheter (Abbott Inc., Illinois, USA), was used to map and tag conduction
system potentials within the sub-pulmonary LV (Figure 1).

The HD grid has a relatively large footprint (13x13mm), but can be compressed to fit
through an 8.5Fr sheath. To facilitate direct delivery of the HD grid into the sub-pulmonary
LV, and avoid damage to the peripheral veins or the newly implanted (and thus non-
endothelialised) SVC stent, the 8Fr short sheath was exchanged for an 8.5Fr SLO sheath (St
Jude Medical, Minnesota, USA) which was advanced over the guidewire leaving the tip of
the sheath just beyond the SVC stent. In addition to the 3D electroanatomical map,
fluoroscopic images of the HD grid, positioned over conduction system potentials, were
taken in orthogonal views (LAO 30 and RAO 30) to act as a second reference. Once a
suitable left bundle target was identified, a standard left-sided infraclavicular incision was
made and axillary vein access obtained under ultrasound guidance. A C135 sheath
(Medtronic, Minneapolis, USA) was manually reshaped (Supplementary Video 1) to the
atrial switch anatomy of the patient by reversing the direction of the primary curve and
augmenting the secondary curve (Figure 2). With the sheath modified in this fashion,
clockwise torque by the operator, once in the LV, places the tip of the sheath against the
interventricular septum.

The distal pole of a 3830 SelectSecure lead (Medtronic, Minneapolis, USA) was
connected in a unipolar fashion to the mapping system to facilitate visualisation of the lead
tip, allowing it to be guided into the conduction system via the modified C315 sheath. The
position was confirmed fluoroscopically before deploying the lead (Figure 3).

In contrast to the traditional placement of the pacing lead in left bundle area (where
both electrodes are located within the septum), the lead was tunnelled so that just the distal
electrode was in the septum due to the relatively superficial location of the left bundle system
when approached from the morphological left ventricle. A second lead was deployed in the
left atrial appendage.

Electrical parameters were excellent with left bundle capture threshold at
1.0V@1.0ms. The resultant QRS duration (142ms) and morphology were almost identical to
that of the intrinsic QRS.

Discussion:
Systemic RV dysfunction, frequently exacerbated by LV apical pacing, is an
important cause of morbidity and mortality in this population\textsuperscript{4}. It is accepted that CRT is
beneficial and possible, but requires an epicardial or hybrid transvenous/epicardial approach.
Providing physiological pacing via a completely transvenous approach avoids the additional
risk of cardiothoracic surgery. Cano et al have demonstrated with feasibility of conduction
system pacing in ACHD, our use of visualisation of the pacing lead through the
electroanatomical mapping system allows for directly guided lead placement into the left
bundle\textsuperscript{10}. 
In TGA the conduction system is located in the traditional anatomical location within the ventricles in a similar distribution to that in usual cardiac anatomy. Complexity arises from the need to deliver the left bundle lead from the left ventricle when the implant sheaths are designed for an RV approach to the septum. The C135 sheath is designed for delivery of a 3830 lead onto the His-bundle, and more recently has been used for implantation in the septum for left-bundle area pacing. It has a primary curve designed to angle the sheath rightwards (when viewed in an AP projection) through the RA and tricuspid valve, and a secondary curve at the distal tip with a 90-degree offset allowing for septal (posterior) angulation. In patients with TGA and atrial switch, the sub-pulmonary LV is posterior relative to the systemic RV; without modification, the C315 sheath would be directed towards the posterolateral LV wall (away from the septal left bundle system). To address this, we re-shaped the primary curve of the C315 sheath manually such that it was reversed by 180 degrees. The catheter can then be ‘flipped’ – thus retaining the primary angulation, but reversing the secondary curve to face anteriorly towards the septum (Video 1). It is important to perform this manoeuvre with the dilator in-situ to prevent deformation of the lumen of the catheter which may subsequently impede torque transmission during lead deployment.

Adding further complexity to the desired shape is that the anterior leaflet of the mitral valve opens towards the septum and must be circumnavigated – necessitating a much deeper primary curve and sharper distal curve. This is not an issue on the right side, as the lead deployment location approximates the raphe between the septal and posterior leaflets of the tricuspid valve. Re-shaping the C315 sheath is a well-known and used technique to achieve His-bundle and left-bundle pacing in usual cardiac anatomy, but the adjustments required to the sheath shape are minimal in these instances\textsuperscript{11}. In our case the adjustment was more radical with complete reversal of the primary curve; however, we found the sheath re-shaping to be well tolerated by the sheath and the new shape retained for the duration of the implant.
Left bundle pacing provides synchronous activation of the left ventricle, but the long-term effects of the resultant RBBB has not been studied. Spontaneous, lone RBBB has no adverse effect in the general population, but in patients with a systemic RV the effect of the resultant RBBB is unknown. Sub-pulmonary LV dysfunction is associated with an increased incidence of clinical heart failure and biventricular CRT has been shown to improve systemic RV function. There is a need for collaboration between groups to establish if conduction system pacing has a similar beneficial effect and what role it has in the future of ACHD pacing.

Conclusions:

Conduction system pacing in atrial switch TGA patients is feasible, maintains intrinsic ventricular depolarisation, and may provide improved long-term outcomes regarding systemic RV function. Implant tools currently available require substantive adaptation to adjust for patient-specific anatomy, and the use of electroanatomical mapping is helpful to guide the implant. Given the heterogeneity of anatomy in complex ACHD patients, further study is warranted to confirm reproducibility of this technique and correlate with physiological and clinical outcomes.

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References:


Figure 1 Electroanatomical map (RAO) with conduction tissue tagged (yellow) and final lead position (green) highlighted. EGMs from specific locations demonstrate progressive shortening of the ‘HV interval’ with more distal conduction tissue sampling.

Figure 2 (A) An original C315 Sheath (left) and the reshaped sheath (right) to adapt for the Mustard anatomy with a deep primary curve under the mitral valve and an anterior or right-hand secondary curve to the septum. (B) The reshaped C315 sheath in an explanted Mustard heart with the sub-pulmonary left ventricular free wall reflected to demonstrate the different shape required to reach the left bundle. Dashed lines outline the approximate location of the proximal left bundle conduction system. * marks the anterior mitral valve leaflet.

Figure 3 (A) Fluoroscopic image demonstrating SVC and IVC baffles and final lead position with reference to the HD grid catheter. (B) Presenting ECG demonstrating sinus rhythm, complete AV block and a narrow complex escape rhythm. (C) Final paced ECG demonstrating left bundle capture with almost identical QRS morphology to the intrinsic QRS.
Figure 1
Figure 2
Figure 3
Key Teaching Points

1. Left bundle pacing in complex ACHD anatomy such as transposition of great arteries post-atrial switch is feasible with potential benefits.
2. Left bundle pacing can be facilitated by the use of 3D electroanatomical mapping with visualisation of the pacing lead.
3. The Medtronic C315 sheath can be extensively re-shaped and adapted to suit complex anatomies; doing so with the dilator in situ is important to prevent damage to the sheath.