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Permanent Pacemaker Implantation in an Adult with a Rare Congenital Anomaly and Dilated Cardiomyopathy

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NADİR BİR DOĞUŞTAN ANOMALİ ve DİLATE KARDİYOMİYOPATİLİ BİR ERİŞKİNDE KALICI PACEMAKER YERLEŞTİRİLMESİ

ÖZET

66 yaşında dekstrokardili erkek hasta, dilate kardiomiyopatiye bağlı konjestif kalp yetersizliği ve eşlik eden semptomatik bradiaritmi nedeni ile kalıcı kalp pili uygulanmak üzere yatırıldı. Kalıcı kalp pili takılması sırasında karşılaşılan güçlük üzerine pek çok kez verilen kontrast madde enjeksiyonları ile sol taraftan seyirli inferior vena cavanın karaciğer düzeyinde kesintiye uğradığı ve azygos devamlılığı göstererek anormal şekilli sağ atriuma girmeden önce kangal oluşturduğu, sağ tarafta persistan vena cava superiorun bulunduğu ve hepatik venlerin sağ atriuma alttan bağlandığı gösterildi. Kalıcı kalp pili daha sonra başarı ile yerleştirildi. Nadir konjenital anomalili hastalarda transvenöz kalıcı kalp pili uygulaması öncesi kontrast madde enjeksiyonu ile sağ ventrikül apeks yerinin tam olarak belirlenmesi oldukça önemlidir.

Anahtar kelimeler: *Kalıcı kalp pili-dekstrokardi-persistan superior vena cava-inferior vena cava*

Dilated cardiomyopathy(DCMP) is a common outcome of adult congenital heart disease of various forms. Bradyarrythmias, presumably secondary to AV conducting system degeneration occasionally accompany some of these cardiomyopathies.

CASE REPORT

The patient, 66 year old male, with existing diagnosis of dextrocardia and DCMP for 10 years presented with increasing shortness of breath, orthopnea and dizzy spells over the preceding 5 months. His previous medications included indapamide, digoxin and atenolol, which were changed to currently taken fosinopril, isosorbid mononitrat, furosemid and colchicine 3 months ago for increasing symptoms. Digoxin was then held for a rather recent onset of persistently low ventricular rate associated with chronic atrial fibrillation. He had had a cardiac catheterisation 10 years ago with normal coronaries and diffuse, severely hypokinetic and enlarged left ventricle(LV). He also had dextrocardia with situs inversus and no other associated abnormality was mentioned. On physical examination, he was in mild respiratory distress. His blood pressure was 100/63 mmHg and heart rate was 40/min. His neck veins were elevated, 5-6 cm, at 45 degree. He had a sustained and displaced apical impulse at right precordium, end inspiratory crackles in both hemithorax bases and +2 oedema in both legs. The admission hemogram was normal, blood creatinin was 2.2 mg/dl and K was 4.3 mmol/1. His ECG showed atrial fibrillation, ventricular rate of 30-40/min, extreme right axis deviation,QRS duration of 200 ms and QS complexes in V1-V6.Admission chest X-ray showed a right sided cardiac silhouette with increased cardiothoracic ratio and clear lung fields. Echocardiogram showed situs inversus dextrocardia with 4 chamber enlargement, with mild tricuspid regurgitation(TR) (peak TR jet velocity 3.4 m/sec) and mild mitral regurgitation. A right-sided cardiac catheterisation done prior to pacemaker implantation revealed the most unusual findings. The right groin was prepared and 7F sheath was inserted into femoral vein under sterile conditions, then a 6F pigtail catheter was advanced to inferior vena cava (IVC) and into right atrium over a very unusual course (Fig 1). Interruption of IVC with left sided azygos continuity which made a loop in thorax before entering into right atrium was demonstrated with repeat contrast injections as well as a very enlarged and abnormally shaped right atrium with a right sided persistent superior vena cava (SVC). There was no de-

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Figure 1. A very unusual course of a pigtail catheter advanced to right atrium via azygos continuity of inferior vena cava which made a loop in thorax before entering into right atrium has been demonstrated.

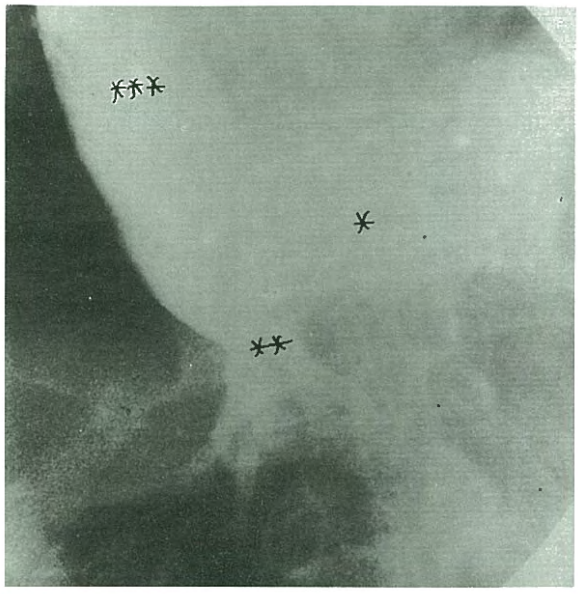

Figure 2. A very enlarged and abnormally shaped right atrium with a right sided superior vena cava and hepatic veins connecting to the inferior portion of right atrium can be seen.
* right atrium * ** hepatic veins * ***right sided superior vena cava

monstrable left sided SVC or other communication with the right sided SVC. Hepatic veins were seen to connect directly and separately to atria from below (Fig 2). A high fidelity measurement indicated a mean pressure of 13 mmHg in right atrium. After repeated attempts catheter was passed into right ventricle and a right ventriculography was done (Fig 3a).

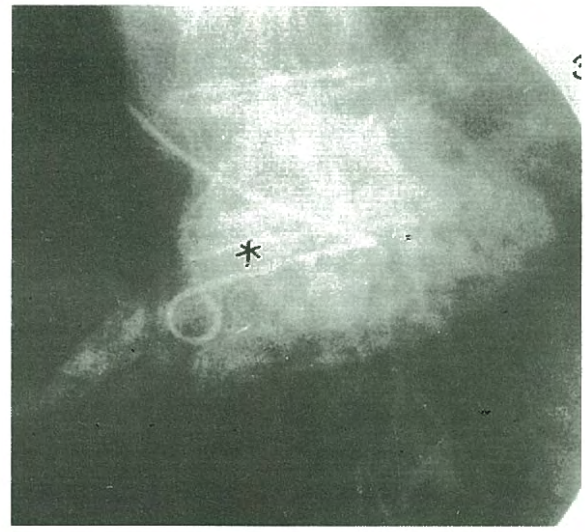

Figure 3. A WI Pacemaker (telectronics) was placed into right ventricular apex via left subclavian vein successfully.

Again a high fidelity measurement indicated a right ventricular(RV) pressure of 100/36 mmHg. The catheter could not be further advanced to the main pulmonary artery. The next session, a WI Pacemaker (Telectronics, model no: 8218) was placed into right ventricular apex via left subclavian vein after several attempts (Fig 3b). The technical difficulty rose from abnormally low location of right atrioventricular groove and tricuspid valve. The final ventricular stimulus threshold was 1.2 V, lead impedance was 420 ohms and lower rate was set at 60/min.

DISCUSSION

The case reported herein underscores the potential problems and pitfalls encountered during permanent pacemaker placements in patients with congenital heart disease and also describes a very unique association of an extremely unusual systemic venous return abnormality and situs inversus type dextrocardia. The unusual course of NC in our patient possibly represent a manifestation of visceral heterotaxy and left atrial isomerism (LAI).It has been reported that hepatic veins which were shown to connect directly to atria from below were invariably associated with LAI [1].The azygos extension of an interrupted IVC leading to SVC in LAI has been well defined [2]. The unusual looping configuration of azygos extension in our patient may be the result of an atretic left sided SVC [3]. We presume that the originality

of current report is that it is the first reported case about pacemaker implantation in such a particularly unusual congenital cardiac complex in an adult. There has been a report about heart block in dextrocardia with situs inversus stating that this case represented a very rare association [4]. Another report was about permanent cardiac stimulation in a patient with isolated dextrocardia and ventricular septal defect [5]. Only 6 cases among 12 days to 13 years old children with left isomerism and complete AV block all of whom required pacemaker implantation with only one surviving after the procedure, have been reported [6].

As has been mentioned in this case report, precise knowledge of the venous system and the location of the apex of the right ventricle is necessary prior to permanent pacemaker implantation.

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