行政院國家科學委員會專題研究計畫成果報告

Preparation of NSC Project Reports

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一、中文摘要

先天心臟病病兒常合併心傳導系統異常。這些傳導異常性質可能導致臨床不整脈，進而影響其預後。此外，經開心手術成功矯治後的先天性心臟病兒，由於手術瘢痕等問題易有心房或心室繞行性頻脈，藥物治療相當困難。

本研究探討先天性心臟病術前術後之心律不整，(1)其型態以其特殊之機轉(2)以傳統心電生理檢查方法及電燒灼術治療之可行性，以及（3）以 Electroanatomical mapping （CARTO system）輔助之不整脈定位及燒灼術之適應性。由 1993 年到 2001 年，共 33 位接受心電生理檢查病兒（占所有病兒 10%）合併有先天性心臟病。有三位病兒之頻脈必須採用 CARTO system 輔助之定位與燒灼（二例心房繞行頻脈，一例心室繞行頻脈）。在術前之病兒，兩側右心房症病兒有相當高的概率合併一種相當特殊的心頻脈，我們稱之為 AV nodal-to-AV nodal tadycardia。反之，兩側左心房症病兒易有竇房結及房室結功能不良。此外，先天矯正型大血管轉位病也有常有上心室頻脈。這些術前心律不整大多可以傳統之電生理檢查方法與電燒灼術等予以根治，然而先天性心臟病兒術後之頻脈若與開刀瘢痕有關，則復發率高，且常須輔以須輔以 CARTO system。但即使如此，復發率仍偏高。

CARTO system 由於尺寸較大，不適用於較小兒童，且其導管價格極為昂貴。因此，較適用於開心手術後繞行性頻脈之定位與燒灼。

關鍵詞：先天性心臟病、心律不整

Abstract

Background. Congenital heart disease may be associated with conduction system abnormalities that may lead to clinical arrhythmias and significant morbidity.
Methods. Patients with congenital heart disease who had clinical arrhythmias were enrolled. Standard electrophysiological study and mandatory interventions, including radiofrequency ablation and pacemaker implantation, were performed under propofol anesthesia. Electroanatomical mapping were used when standard electrophysiological study failed to identify or eliminate theclinical arrhythmias.
Results. From 1993 to 2001, a total of 33 out of 317 (10%) patients received standard electrophysiological study for clinical arrhythmias were found to be associated with congenital heart disease. Carto system (electroanatomical mapping) was required in three patients, due to postoperative reentrant atrial tachycardia in two and reentrant ventricular tachycardia in one.
In preoperative evaluation, the right atrial isomerism was closely associated with supraventricular tachycardia that has been characterized as AV nodal-to-AV nodal tachycardia. In contrast, patients with left atrial isomerism were highly possible to develop sick sinus syndrome and AV conduction disturbance. Patients with corrected transposition of great arteries could also be associated with atrial flutter, atrial tachycardia or AV nodal reentrant tachycardia. Postoperative cardiac patients could develop reentrant tachycardia attributed to accessory pathways or incisional atrial or ventricular reentrant tachycardia. The incisional reentrant tachycardia carried high recurrence risks even using the CARTO

system

Conclusion.

Patients with certain types of congenital heart diseases, including heterotaxy syndrome and corrected transposition of great arteries tended to have clinical arrhythmias during childhood and should be managed before surgical intervention. Postoperative incisional reentrant tachycardia carried high recurrence rate even with CARTO system assisted RF ablation.

Keywords: Congenital heart disease, tachycardia, bradycardia, electroanatomical mapping

二、緣由與目的

The incidence of congenital heart disease (CHD) is 8/1,000 live birth and remains relatively constant.[1] Over the last few decades the prognosis of children with CHD has been improved, primarily due to new catheter or surgical interventions that fundamentally changed the clinical course, enhanced quality-of-life and prolonged life expectancy. In Taiwan, the first success surgical repair of cardiac defect was performed in 1964 and first catheter palliation of pulmonary stenosis was done in 1986. In recent 5 years, the case numbers and complexities of catheter and surgical interventions for CHD increased rapidly. Therefore, an increasing number of patients survive into late childhood. Although most of the patients may have satisfactory hemodynamics, a substantial of patients develop cardiac arrhythmias during the long-term follow-up. [2-3] The etiologies may include 1) abnormal conduction system associated with the complex congenital heart disease, 2) dilated cardiac chambers associated with unsatisfactory hemodynamics, 3) altered ultrastructure in chronic abnormal load, 4) multiple surgical scars after staged operation, etc.[4-6] The development of cardiac arrhythmias frequently complicates the clinical course and may leads to significant morbidity and mortality. Nonetheless, the related studies are still limited and most of them addressed the unsatisfactory results of current treatments. The poor results are related to underlying complex cardiac defects and surgical scars, which are difficult to be delineated by conventional electrophysiological study methods

Drug therapy for the arrhythmias found in cardiac children is usually unsatisfactory and was often associated with negative inotropic effects which may worsen the borderline hemodynamics of cardiac children.[7] The application of radiofrequency catheter ablation offers the opportunity to cure the arrhythmias.[8] Currently, the mapping/ablation study is performed by using multichannel recording technique and under fluoroscopy guidance. Its efficacy has been well shown in terminating the atrioventricular reentrant tachycardia, atrioventricular nodal reentrant tachycardia, atrial flutter and ventricular tachycardia.[8-10] However, its efficacy in eliminating the substracts supporting the tachycardia in postoperative cardiac children is still limited.[11-12] The major drawbacks are: 1) the complex cardiac anatomy found in postoperative cardiac children is difficult to be delineate by using the biplane fluoroscopy, 2) the multiple scars and surgical suture load of previous palliative surgery may be associated with complex multiple reentrant loops which is difficult to be defined by biplane fluoroscope, 3) the endocardial surface is invisible using the fluoroscopy and the target sites can only be approximated by their relationship with nearby structures such as ribs, blood vessels and the position of other catheters, and 4) the irradiation exposure on the growing children. The recent development of a novel system, nonfluoroscopic mapping/ablation system (CARTO system) may help to solve the problems.[13]

The most common postoperative arrhythmia is intratrial reentrant tcahycardia, resulting from circus movement of activation wave fronts around either surgical incisions and/or other anatomical conduction barriers such as conduit, patches for closure of defects, orifices of the great veins, and the atrioventricular annulus.[11-12] These anatomical barriers prevent the lateral

collision of reentrant wave front and form the underlying substracts for reentrant tachycardia. It occurs as commonly as in 30% of patients after atrial switch operation (Mustard/Senning procedure for transposition of the great arteries) and in 50-60% of patients after palliative Fontan operation for single ventricle physiology at 5-year follow-up.[14-15] In Taiwan, the first Mustard operation was performed in 1974. Although the atrial switch operation had been gradually replaced by arterial switch operation, the patients with previous atrial switch operation for their transposition of great arteries still constitute significant patient population. There are also increasing number of patients receiving palliative Fontan type circulation, such as tricuspid atresia, single ventricle and right atrial isomerism complex. We are facing increasing numbers of GUCH (grown-up congenital heart) patients with arrhyuthmias. The complex cardiac conduction system found in patients with right atrial isomerism further increases the complexity of the arrhythmias. [16-17]

In our center, we performed the first conventional RF catheter ablation in children in 1994. The results of the first five-year (1994 to 1998) have shown that the overall successful RF ablation for all types of arrhythmias was 96%. [18] The accessary pathways, and dual AV nodal pathway could be successfully treated in almost every case. But the results in postoperative arrhythmias, as reported by other centers, were still limited. The successful rate was only 57% and with recurrence in 25% of the successful cases. To resolve such tuff issue, the potential of electroanatomic mapping/ablation system is addressed.[13,19-22] The nonfluroscopic electroanatomic mapping/ablation system (CARTO system) represents a paradigm shift in the ability to map the three-dimensional anatomy of the heart and determines the cardiac electrical activity at any given mapped point.[13] The feasibility of such mapping/ablation system have been well demonstrated in some recent studies, including those with accessary pathways, atrial flutter, atrial fibrillation and ventricular tachycardia.[19-23] The potential benefits have also been suggested in limited studiesof postoperative congenital heart disease.[24]

三、結果與討論

RESULTS

Patients

From 1993 to 2001, a total of 33 out of 317 (10%) patients received standard electrophysiological study for clinical arrhythmias were found to be associated with congenital heart disease. Carto system (electroanatomical mappi ng) was required in three patients, due to postoperative reentrant atrial tachycardia in two and reentrant ventricular tachycardia in one.

In preoperative evaluation, the right atrial isomerism was closely associated with supraventricular tachycardia that has been characterized as AV nodal-to-AV nodal tachycardia (7/7, successful RFCA using conventional EPS modalities 3/3). In contrast, patients with left atrial isomerism were highly possible to develop sick sinus syndrome and AV conduction disturbance at follow-up (5/6). One received DDD pacemaker implantation at the age of 5 years. Patients with corrected transposition of great arteries could also be associated with atrial flutter, atrial tachycardia or AV nodal reentrant tachycardia. Initial success was obtained in all patients with sustained tachycardia.

Postoperative cardiac patients could develop reentrant tachycardia attributed to accessory pathways (2) or incisional atrial (10) or ventricular reentrant tachycardia (2). Although initial success may be achieved in 13 patients, three patients required the CARTO-assisted RFCA to eliminate the tachycardia. But, the recurrence rate (2/3, 67%) was still high.

The major drawbacks of using the CARTO system for mapping and ablation of difficult arrhythmias in children were the size (7F, 8F sheath) and the cost of the catheters. Nonetheless, this system may still can be useful for children with postoperative incisional tachycardia that failed to be

eliminated or recurred after conventional study and ablation.

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※　複雜性先天心臟病術前術後心律不整之電生理學　　※
※　機轉：以同步電位及立體結構定位法研究　　　　　※
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計畫類別：□個別型計畫　　□整合型計畫

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中　華　民　國　91　年　10　月　31　日