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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(1/3)

Cardiac Arrhythmias in Patients with Complex Congenital Heart Disease Before and After Surgical Palliations

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First year: Cardiac Rhythm Disturbances in Complex Congenital Heart Disease

Rhythm disturbances in patients with left atrial isomerism

ABSTRACT

Objectives. To determine the prevalence and the electrophysiological mechanisms of rhythm disturbances in patients with left atrial isomerism (LAI).

Background. Defective sinus node and atrioventricular conduction tissue have been described in the hearts associated with LAI.

Methods. From 1984 to 1998, a total of 22 patients, and from 1995 to 1998, 3 fetuses, were identified as LAI and constituted the study population. Pathological confirmation was obtained in 14 patients and 1 fetus.

Results. The age at the last follow-up ranged from 2 to 276 months (90± 70 months). Three fetuses (100%) developed sinus bradycardia and intermittent AV block, and were ended with heart failure and termination. Associated cardiovascular anomalies of the 22 pediatric patients were interruption of inferior vena cava (18, 82%), common atrium (9, 41%), AV canal (14, 64%), double-outlet right ventricle (8, 36%) and pulmonary stenosis (15, 68%). Over half of the patients (14, 64%) developed bradyarrhythmia (onset age, 1 to 264 months, median 78 months): junctional escape rhythm as dominant rhythm, 10 (45%), sinus bradycardia or sinoatrial block, 8 (35%, 5 of the 7 also had dominant junctional rhythm) and AV block, 2 (9%, both had dominant junctional rhythm). By actuarial analysis, the probability free from bradyarrhythmia decreased with age and was 80% and 46% at the age of 2 and 6 years, respectively. Two patients experienced fainting attacks. However, junctional ectopic tachycardia after the cardiac operation occurred in 3 out of 10 patients. Electrophysiological studies (3 cases) revealed sinus node dysfunction in 2/2 and impaired AV conduction in 1/2. Besides, a Mahaim-like pathway (progressive shortening of HV interval during decremental atrial pacing) was identified in both of the patients with His potential recorded during the study.

Conclusions. Over half of the LAI patients may develop bradyarrhythmia at the age of 6 years mainly due to the subnormal sinus node function. Bradyarrhythmia appeared during the fetal stage may cause preferential loss of the fetuses. Patients with LAI also have a higher chance to have junctional tachycardia which may be related to the abnormal AV conduction properties. The high prevalence of Mahaim-like pathway warrants further investigation.

Keyword: left atrial isomerism, heterotaxy syndrome, sinus bradycardia, junctional rhythm, junctional tachycardia

Left atrial isomerism (LAI), a form of heterotaxy syndrome, is characterized by the bilateral left atrial morphology of the atria as well as the interruption of inferior vena cava with azygous continuation (1,2). Intracradiac anomalies may be present, but the severity is usually less severe than those found in patients with right atrial isomerism (1-4). Associated intracardiac anomalies usually include partial anomalous pulmonary venous return, common atrium, atrioventricular (AV) canal, double-outlet right ventricle and pulmonary stenosis, although the incidence of double-outlet right ventricle and pulmonary stenosis was much lower than that found in right atrial isomerism. As to the conduction system, the sinus node has been described as defective, hypoplasic or even absent (5,6). The AV nodes may be single or paired, but is frequently associated with a discontinuity between the AV node and the ventricular conduction tissues. Such conduction system may result in sinus node dysfunction or AV block (7). Atrioventricular block had been shown in 15 % of patients and nodal rhythm in 12% of the patients with LAI (7). Since most of the patients are associated with complex congenital heart disease, the long-term prognosis was deemed as determined by the associated cardiac anomalies and only rarely permanent pacemaker was implanted. However, the recent advances in surgical or transcatheter palliations for complex cardiac lesions have improved the long-term outcome. Therefore, it becomes feasible and mandatory to define more clearly the clinical significance of such defective conduction system in LAI patients during the long-term follow-up. This longitudinal study on 22 LAI pediatric patients and 3 LAI fetuses sought to determine the prevalence and the electrophysiological mechanism of the rhythm disturbances in LAI patients.

METHODS

Study patients. From January 1984 to December 1998, a total of 148 pediatric patients were

diagnosed to have heterotaxy syndrome at this institution. From January 1995 to December 1998, by fetal echocardiography 28 fetuses were found to have heterotaxy syndrome. Among them, 22 pediatric patients (8 male and 14 female) and 3 fetuses were LAI and constituted the study population. The diagnosis of LAI was based on a combination of echocardiography, cardiac catheterization/angiography and available magnetic resonance imaging or computerized tomography (8-11). For fetus study, the diagnosis was obtained by fetal echocardiography. Open heart surgery in 14 pediatric patients and autopsy in 1 fetus confirmed the diagnosis. Patients with LAI have 1) bilateral finger-like atrial appendages, 2) bilateral hyparterial bronchi, or 3) interruption of inferior vena cava with azygous continuation and with the cross section of azygous vein posterior to the descending aorta at the level of T_{10}. The diagnosis of rhythm disturbances was based on the serial 12-lead electrocardiogram. The rhythm found during awake time and persisted in the next follow-up EKG was defined as dominant rhythm. The diagnosis of cardiac rhythm in fetuses was based on the fetal echocardiography.

Statistics

Data were expressed as mean± SD. Actuarial event-free curves were drawn according to the nonparametric estimation by Kaplan and Meier (12). *Chi-square* test was used to examine the significance when appropriate.

RESULTS

Demographics

Three fetuses were all ended with termination before the 26 gestation weeks. Common atrium, complete AV canal and interruption of inferior vena cava with azygous continuation were identified in all. Excluding the 3 fetuses, the age at the last follow-up ranged from 2 to 276

months (90± 70 months. median 59 months). The associated cardiac anomalies were summarized in Table 1. Interruption of the inferior vena cava was found in most of the patients. The association of AV canal and common atrium was also high. One patient was found to have no significant intracardiac anomalies. The actuarial analysis of survival of the patients revealed a ten-year survival of 70% (Fig. 1). Palliative interventions had been performed in 17 patients (Table 2). The palliations were performed to increase the pulmonary flow in 7, to repair the septal defect in 4, and to reach a Fontan type circulation in 6.

Rhythm disturbances

All three fetuses developed sinus bradycardia and intermittent AV block before the 26 weeks' gestation. Hydrops fetalis developed in 2 cases. Sinus rhythm either from the right-sided LA or left-sided LA was noted in 14 patients at initial presentation, and 6 patients had low atrial rhythm. During the follow-up, over half of the patients (14/22, 64%) developed bradyarrhythmia. Junctional escape rhythm as dominant rhythm (varied from 46 to 89/min, 62± 13/min, median 60/min), in 10 (45%), sinus bradycardia or sinoatrial block, in 8 (35%, 5 of the 7 also had dominant junctional escape rhythm) and AV block in, 2 (9%, both had dominant junctional rhythm). The onset age of the bradyarrhythmia ranged from 1 month to 264 months (75±85 months, median 78 months). By actuarial analysis, the probability being free from bardyarrhythmias was 80% and 46% at the age of 2 and 6 years, respectively (Fig. 2). However, only two patients experienced fainting attacks. None of the patients had bradyarrhythmia directly related morbidity and mortality. None had received permanent pacemaker implantation.

Three patients developed junctional ectopic tachycardia after open heart surgery, and one of them also had junctional ectopic tachycardia during the rapid atrial pacing of the

electrophysiological study. One of them had sinus pause detected during Holtor monitoring, and none of the patients before the development of junctional ectopic tachycardia had junctional escape rhythm as the dominant rhythm. The juctional ectopic tachycardia varied with a heart rate from 120 to 215/min and was associated with unstable hemodynamics. The tachycardia didn't respond to verapamil, digoxin, proprnolol or cardioversion, and was converted to sinus or low atrial rhythm only after hemodynamics improved by cardiopulmonary resuscitation in two. Adenosine had been used in one patient and might temporarily slowed down the rate of tachycardia. One patient had right heart failure after total cavopulmonary connection operation. He suddenly developed junctional ectopic tachycardia (maximal rate 215/min) 4 months after the operation. His condition deteriorated and died on the same day. As compared to the other 7 patients who had also received intracardiac surgery but without postoperative junctional ectopic tachycardia, we found that the none of the clinical characteristics, including gender, age at operation, pre-existing rhythm and ventricular morphology, was associated with a higher risk of developing junctional ectopic tachycardia after interventions.

Electrophysiological study

Electrophysiological study was performed in 3 under propofol anesthesia, but the study was limited by the development of junctional ectopic tachycardia with unstable hemodynamics in one case during rapid atrial pacing. The electrophysiological parameters are summarized in the Table 3. Alternating rhythm from right or left sided left atria was found in 2 and junctional rhythm in one. The sinus node function was abnormal in both patients in whom it had been examined: prolonged maximal corrected sinus recovery time in one and a slow intrinsic heart rate in the other. The sinus node recovery curve (assessed by Narula method) in these two patients showed only nonreset responses, which implied the presence of sinus

node entrance block. The Wenckebach cycle length (the atrial pacing cycle length with loss of 1:1 AV conduction above the His bundle) of the AV conduction was longer than the normal age control in one. Whereas in the other patient, the atrium failed to maintain the heart rate at the pacing cycle length of 300 ms, and therefore limited the study of AV node. By decremental atrial pacing, we found that, in both patients who had clearly recorded His potential, the His potential was moved to the ventricular activation at shorter pacing cycle length (Fig. 3) or even merged into the ventricular activation (Fig.4). The relation between atrial pacing cycle length and HV interval showed progressively shortened HV interval along with lengthening of the AH interval during decremental atrial pacing (Fig. 5). But, changes of the QRS morphology was only evident in one case. Such behavior of AV conduction suggested the presence of Mahaim-like accessory pathways. However, no tachycardia nor echo beats were induced. It is possible that the antegrade AV conduction was through AV node as well as a direct extension from AV node (or His bundle) to fascicular or ventricular tissue. Since we did not map the discrete potential and the insertion of the accessory pathway, we preferred to use the term “Mahaim-like fiber” to indicate the behavior of progressive shortening of the HV interval during decremental atrial pacing. The degree of QRS changes would depend on the closeness between the location of normal AV node conduction axis and the Mahaim-like fibers.

DISCUSSION

Although the pathological description of defective conduction system in LAI patients had been described as early as 1975, the long-term clinical significance remains unclear. This longitudinal study of 22 LAI patients and 3 fetuses have delineated the high probability of developing bradyarrhythmias during the follow-up. But the unique findings of this study

were the propensity to have junctional ectopic tachycardia and the high incidence of Mahaim-like accessory pathways in LAI patients.

Natural History of the Bradyarrhythmia in LAI Patients

Previous reports suggest an incidence of bradyarrhythmias ranged from 20 to 30% in LAI patients (7, 13,14). In this study, the incidence of bradyarrhythmias was as high as 64%. By event free analysis, the probability free from bradyarrhythmia decreased with age and over half of the patients would experience bradyarrhythmia when they reached the age of 6 years. In the series reported by Wren et al (6), bradyarrhythmia at initial presentation was noted in 27% of the patients and by Holtor monitoring in 64% (9/14) of the patients. Since we have performed serial EKG in most of the patients, the incidence of bradyarrhythmia found at our study would be more close to that defined by Holtor. The sinus node in LAI patients is usually abnormal (5,6). As described by Dickinson et al (6), although a sinus node can be identified on the junction of the right-sided or left-sided atrial appendage with the atrium in about half of the patients, the sinus node was abnormally small in all. In about one-fourth of their patients, no sinus node could be identified. The report from Ho et al has disclosed absent sinus node in three-forth of their patients (5). Such pathological evidences may account for the high probability of sinus node dysfunction in LAI patients. Furthermore, the function of the hypoplastic sinus node may deteriorate with time and thereby the probability being free from bradyarrhythmias in these patients will decrease with age. The sinus node dysfunction in LAI patients as shown in this study might be present as dominant junctional escape rhythm, low intrinsic heart rate, prolonged sinus recovery time and sinoatrial entrance block. In the previous report by Wren et al (7), a significant portion (12%) of LAI patients similarly developed nodal rhythm. The incidence of nodal rhythm was even higher as shown by Holtor monitoring. Evidences of sinus pause suggesting sinus node dysfunction had also been

shown in 2 out of the 14 patients receiving Holtor monitoring. As to the AV nodes, four patterns could be summarized from the previous pathological reports: 1) one AV node with preserved connection to atrial transitional cells but lost connection to nonbranch or right bundle, 2) one AV node with normal connection, 3) two AV nodes connected by a sling of conduction tissue (Monckeberg sling) which descends into the bundle branches, and 4) two AV nodes lost connection to the sling (5,6). Such varied AV conduction system may result in a variety of AV conduction disturbances. Previous reports had emphasized the high incidence of AV block in LAI patients and none of the reports mentioned the occurrences of tachycardia. The incidence of AV block ranged from 7 to 30% (7, 13,14). The AV block (3 with first-degree, 2 with second-degree and 5 with complete) was demonstrated in 10 out of 67 patients in the report by Wren et al. (7). Garcia et al (13) found complete AV block in 6 out of 30 patients and Rougin et al (14) reported complete AV block in 3 out of 11 cases. Complete AV block during the fetal life may cause hydrops fetalis and fetal loss (15). In this report, we have only 2 cases (9%) of AV block and both also had dominant junctional rhythm. All fetuses developed bradycardia which had been characterized as sinus bradycardia with intermittent AV block. Based on these observations, we suggest that the bradyarrhythmias found in LAI patients are mainly due to abnormal sinus node function and in some cases the abnormal sinus node may be associated with compromised AV conduction. When the sinus rate is low, the junctional rhythm will appear. However, when the sinus rate is relatively high, then AV block may appear because the compromised AV conduction is unmasked at a higher sinus rate.

Tachyarrhythmia in LAI Patients

In this study we have also noticed a propensity (30%) to develop junctional ectopic tachycardia after cardiac operation in LAI patients. As compared to the incidences of

postoperative junctional ectopic tachycardia after Fontan type operation, the incidence was significantly higher (16). Among the 151 patients reported by Friedman et al, 12 patients (8%) was found to have junctional ectopic tachycardia after the Fontan type operation (16). Although we have identified the Mahaim-like accessory pathway in these patients, the tachycardia was not reentrant tachycardia between the accessory pathway and the AV node. The responses to pacing and drugs suggested an automaticity for the mechanism of tachycardia. The pathological basis for the Mahaim pathway in normal hearts has been described as anatomical connections of the AV node to the myocardial septum as well as connections of the origin of the left bundle branch to the upper part of the interventricular septum (17). However, the results of nonpharmocological therapy for such preexcitation, e.g., radiofrequency ablation or surgical ablation, had suggested that such preexcitation originates from remnants of the specialized AV ring tissue (18,19). The Mahaim pathways may be further classified as atriofascicular, nodofascicular or fasciculoventricular pathway based on the detailed electrophysiological mapping (18). Nonetheless, the anatomic basis for the nodofascicular and fasciculoventricular pathways is still ill-defined. Furthermore, such criteria may be not applicable in the presence of associated complex congenital heart disease. Therefore, based on the findings of progressive shortening of the HV interval during decremental atrial pacing in the two cases we studied, we could only reach the diagnosis of Mahaim-like accessory pathways for such abnormal AV conduction. Progressive QRS changes which depends on the closeness between distal insertion sites of the normal and accessory pathway were only found in one case. In one case, tachycardia with VA dissociation developed during rapid atrial pacing study. This junctional tachycardia with a same QRS morphology of the sinus rhythm was associated with a changing heart rate and was resistant to verapamil, propranolol or adenosine, or programmed pacing. This patient also had

junctional ectopic tachycardia after cardiac operation. Therefore, in LAI patients although the abnormal Mahaim-like accessory pathway may potentially serve as reentrant route for tachycardia, the probability of developing junctional ectopic tachycardia from the abnormal AV conduction tissue is also high as suggested by the results of this study. In contrast, although the pathology of right atrial isomerism shared some similarities with LAI in the presence of AV canal, the presence of such Mahaim-like pathways was not found in RAI patients (20,21). In stead, RAI patients were prone to develop reentrant tachycardia between the paired AV nodes (20,21). The influence of situs laterality on the development of cardiac conduction system was suspected but not defined yet. According to the ring theory for the development of cardiac conduction system, the sinoatrial ring tissue contributes to the development of sinus node and transitional cells around the AV node (22,23). As to the atrioventricular ring, it gives rise to AV nodes and an extension of the conduction bundle through the inlet septum. We suspect that the bilateral left-sidedness of the atrium may be associated with an abnormal evolution of the sinoatrial and atrioventricular ring tissues which results in abnormal sinus node, transitional cells, AV node as well as the extension from the AV node. In another words, the abnormal AV conduction tissue in LAI may not only have abnormal connection to the ventricular tissue but also be defective in the regulation of automaticity. By adrenergic stimulation caused by stress, the AV node will be abnormally speeded up in rate and lead to junctional ectopic tachycardia. However, these speculations need to be verified. Previous report had mentioned an evolution of junctional ectopic tachycardia into complete AV block and the histological findings from patients with junctional ectopic tachycardia showed His bundle degeneration, Purkinje cell like tumor and fibroelastosis (23,24). Therefore, it is highly possible that on going pathological changes process in the abnormal AV conduction tissue may manifest as a compromised AV

conduction as well as junctional ectopic tachycardia. In conclusion, this study defines the high probability of developing bradyarrhythmias in LAI patients due to abnormal sinus node function during the long-term follow-up. Varied AV conduction abnormalities may include compromised AV conduction, junctional ectopic tachycardia after intervention as well as the association of Mahaim-like accessory pathway. These rhythm disturbances may change the long-term prognosis of the LAI patients.

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Figure legends

Figure 1. The actuarial survival curve for the 22 patients with left atrial isomerism. The number in the parentheses indicates the number of the patients stayed in the study at the time-pint. The survival decreased with age. The survival at 5 years and 10 years was 87% and 70% , respectively.

Figure 2. The probability free from bradyarrhythmias in 22 patients with left atrial isomerism. The number in the parentheses indicates the number of the patients stayed in the study at the time-pint. The probability free from bradyarrhythmias decreased with age.

Figure 3. The intracardiac electrocardiograms taken during atrial pacing at 600 ms (left) and 310 ms (right), respectively in a 4 years old girl (case 3 in table 3). The HV interval was much shorter at the pacing cycle length of 310 ms than that at pacing cycle length of 600 ms. The changes of QRS morphology can be identified in lead II (not shown), III (not shown), aVF, V1 and V2 (not shown).

Figure 4. The intracardiac electrocardiograms taken during atrial pacing at 480 ms (left) and 300 ms (right) respectively in another 4 years old girl (case 1 in table 3). The HV interval was much shorter and merged unto the ventricular activation at the pacing cycle length of 300ms than that at pacing cycle length of 480 ms.

Figure 5. The relations between the HV interval and the atrial pacing cycle length of the case 3 (A) and case 1 (B). In both, the HV interval was progressively shortened and the AH interval lengthened when the atrial pacing cycle length decreased decrementally.

Table 1. Intracardiac anomalies in 22 patients with left atrial isomerism

Interruption of IVC	18(82%)
Partial anomalous pulmonary venous return	6(27%)
Common atrium	9(41%)
Atrioventricular canal	14(64%)
Double-outlet RV	8(36%)
Pulmonary stenosis	15(68%)
Mitral atresia/hypoplasia	6(27%)
Coarctation of aorta	1(5%)

Table 2. Palliative interventions in 22 patients with left atrial isomerism

Procedures	No. of Cases	Mid-term survival
Balloon valvoplasty	1	1(100%)
Blalock-Taussing shunt	6	6(100%)*
Repair of AV canal or common atrium	4	3(75%)
Hemi-Fontan	2	2(100%)
Total cavopulmonary connection	4	2(50%)
Aortoplasty of coarctation	1	0(0%)

* One received total cavopulmonay connection 4 years after the shunt but died 4 months after the definite operation.

Table 3.Electrophysiological data in 3 LAI patients

Case/sex	Age (yrs)	Anatomy	Dominant rhythm	Sinus node				AV conduction				
				p axis	MCSRT (ms)	IHR	AES	AERP (ms)	location	WCL (ms)	AVERP	Maha
1/female	4	IVC interruption, CA, CAVC, DORV, PS B-T shunt	Sinus, CL 630ms HLA-LLA 30ms AH 80ms HV 24ms	RLA LLA	ND	ND	ND	ND	post	270	ND	
2/female	4	IVC interruption, CAVC, DORV, PS	Sinus, CL 674ms HLA-LLA 20ms AH 75ms HV 73ms	RLA LLA	332	90	nonreset	170 (S_1S_1 600ms)	post	500	470 (S_1S_1 600ms)	
3/female	4	Dextrocardia, narrow hepatic segment of IVC, CA, CAVV, single ventricle, PS	Junctionl ACL 872ms VCL 725ms HV 48ms	RLA LLA	572	ND	nonreset	300 (S_1S_1 600ms)	post ant	≦300	≦AERP	

Abbreviations: ACL:atrial cycle length, AERP:atrial effective refractory period, AVERP:atrioventricula
CA:common atrium, CAVC:common AV canal, DORV:double-outlet right ventricle, IHR:intrinsic heart ra
tachycardia, LLA:left-sided left atrium, MCSRT:maximal corrected sinus recovery time, PS:pulmonary
cavopulmonary connection, VAWCL:ventriculo-atrial Wenckebach cycle length, VCL:ventricular cycle l

Table 4. Comparions between the partients with and without junctional ec
after intracardiae repair

	with JET (n=3)	without JET (n=7)
Male/Female	1/2	2/4
Age at op (yr)	5.3±1.5	9.2±8.2
Dominant Junctional Rhythm		
	0	4
Ventricular morphology		
2 ventricle	3	5
single ventricle	0	2