Surgery for Congenital Heart Disease

CHD

Congenital and surgically acquired Wolff-Parkinson-White syndrome in patients with tricuspid atresia

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Objectives: There are reports associating an increased incidence of Wolff-Parkinson-White syndrome with tricuspid atresia. Here we report on electrophysiologic studies in patients with tricuspid atresia and Wolff-Parkinson-White syndrome after the Fontan operation. In these patients the atrial arrhythmia often seen in patients undergoing the Fontan operation triggered atrioventricular re-entrant tachycardia or caused life-threatening arrhythmias.

Methods: Five patients with tricuspid atresia after palliation with a modified Fontan operation (atrioinfundibular connections) and Wolff-Parkinson-White syndrome are presented.

Results: Four of these patients had symptomatic paroxysmal orthodromic atrioventricular re-entrant tachycardia and a history of syncope; one of them additionally had atrial flutter with 2:1 conduction to the ventricle. A fifth patient presented with a life-threatening broad-complex tachycardia. In electrophysiologic studies an accessory pathway was localized in the right septal area in 3 patients. In 2 patients the accessory atrioventricular pathways were created by means of surgical intervention, connecting the right atrial appendage to the right ventricular outflow tract. All patients could be managed successfully by means of catheter ablation.

Conclusions: In patients with tricuspid atresia, there are congenital and surgically acquired accessory pathways responsible for the increased rate of Wolff-Parkinson-White syndrome. Both types of accessory pathways can and should be treated by means of catheter ablation because atrial arrhythmia often seen in patients undergoing the Fontan operation can trigger atrioventricular re-entrant tachycardia or cause life-threatening tachycardia. Congenital accessory pathways should be excluded carefully before surgical intervention for total cavopulmonary anastomosis in patients with tricuspid atresia.

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Patients after a Fontan procedure are prone to supraventricular arrhythmias.[1,2] Scarring and dilatation of the right ventricle and injury to the sinus node artery contribute to the development of these arrhythmias. Medical treatment, catheter ablation, or both are challenging.

On the other hand, there are several reports of Wolff-Parkinson-White (WPW) syndrome associated with tricuspid atresia.[3-8] Accessory pathways can be localized

TABLE 1. History of the 5 patients with WPW syndrome and tricuspid atresia

	Patient no.				
	1	2	3	4	5
Age at EPS (y)	13	13	14	15	31
Sex	F	F	M	F	M
Operation before Fontan	None	mBTA left	VSD enlargement	VSD enlargement	BTA left, BTA right
Age at Fontan operation (y)	0.5	4	2	0.5	13
Surgical technique					
Fontan anastomosis	Right atrial appendage to right ventricular infundibulum				
Anastomosis patch material	No	*	*	*	*
Closure of the VSD	No	Patch	Patch	Patch	Patch
Division of the RV	Direct suture	No	No	No	No
Recent hemodynamic results					
Systemic CI ($L \cdot min^{-1} \cdot m^{-2}$)	2.8	3.2	2.2	–	1.3
Central venous pressure (mm Hg)	14	10	11	–	8
Pulmonary wedge pressure (mm Hg)	9	5	7	–	3
Arterial oxygen saturation (%)	98	96	93	–	97

EPS, Electrophysiologic study; *mBTA*, modified Blalock-Taussig anastomosis; *BTA*, Blalock-Taussig anastomosis; *VSD*, ventricular septal defect; *RV*, right ventricle; *CI*, cardiac index. *After U-shaped incision of the right atrial appendage, a door-like flap was directly sutured to an incision in the right ventricular infundibulum. The frontal roof of this tunnel-like connection was built with the aid of a polytetrafluoroethylene patch (Gore-Tex patch; W. L. Gore & Associates, Inc, Elkton, Md).

by means of electrophysiologic studies (EPSs) and permanently treated with catheter ablation, even in small children.[9]

We report on the results of EPSs in 5 patients with WPW syndrome and tricuspid atresia after the Fontan-Björk procedure with hemodynamically symptomatic atrioventricular (AV) re-entrant tachycardia.

Patients and Methods

The database of patients with congenital heart disease in our institution listed 198 patients with tricuspid atresia, of whom 16 patients had a suspected antegrade conducting accessory pathway according to surface electrocardiographic (ECG) criteria. Two of these 16 patients died, one as a result of re-entrant tachycardia and another from Glenn thrombosis. Both were 15 years old. Of the remaining 14 surviving patients, 5 agreed to have an EPS performed.

The patients' detailed histories and surface ECG findings before EPS are presented in Tables 1 and 2. All of them had tricuspid atresia with concordant ventriculoarterial connection and reduced blood flow to the lungs (type 1b) caused by a diminutive ventricular septal defect. They were palliated with an atrioinfundibular anastomosis and closure of the connection of the right ventricular infundibulum to the systemic ventricle similar to the description of Björk and colleagues.[10] The hemodynamic results of this operation depicted as recent cardiac catheterization data are shown in Table 1.

Results

EPS was performed under sedation with midazolam. Accessory pathways were localized by means of conventional

TABLE 2. Characteristics of the accessory pathways in surface 12-lead electrocardiography in the 5 patients with WPW syndrome and tricuspid atresia before the electrophysiologic studies

	Patient no.				
	1	2	3	4	5
First occurrence	After Fontan	Before Fontan	After Fontan	After Fontan	After Fontan
PR interval (ms)	80	100	100	60	120
Delta wave negative in lead	avR, V_{1-2}	III, avR, avF, V_{1-2}	III, avR, V_{1-2}	avR, V_{1-2}	II, III, avR, avF, V_{1-2}
Type of clinical tachycardia	*	oAVRT	oAVRT	†	oAVRT
No. of tachycardia episodes	Very rare	Daily	Daily	Weekly	Daily
Antiarrhythmic drug trials	0	9	3	1	4
Predominant rhythm	SR	SR	SR	SR	SR

II, III, avR, avF, V_{1-2}, Standard electrocardiographic surface leads; *oAVRT*, orthodromic atrioventricular re-entrant tachycardia; *SR*, sinus rhythm. *Life-threatening narrow complex tachycardia with a rate of 300 beats/min. †Orthodromic atrioventricular re-entrant tachycardia and atrial fibrillation with 1:2 conduction.

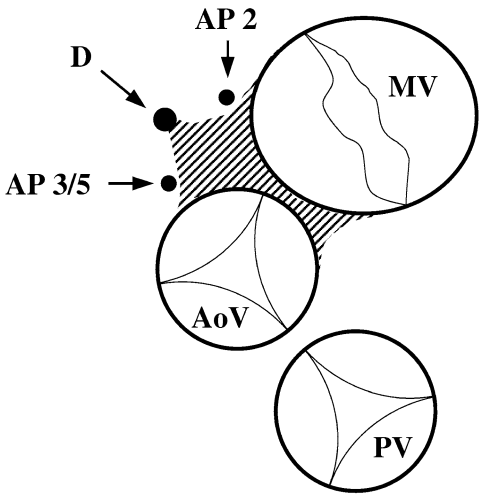

Figure 1. Location of the congenital accessory pathways of patients 2, 3, and 5 depicted schematically on the valvular plane. *MV*, Mitral valve; *AoV*, aortic valve, *PV*, pulmonary valve; *D*, dimple seen in the right atrium just anterior the coronary sinus ostium at the theoretic site of the atretic tricuspid valve, location of the AV node; *AP 2/3/5*, accessory pathway in patients 2, 3, and 5, respectively; *streaked area*, central fibrous body.

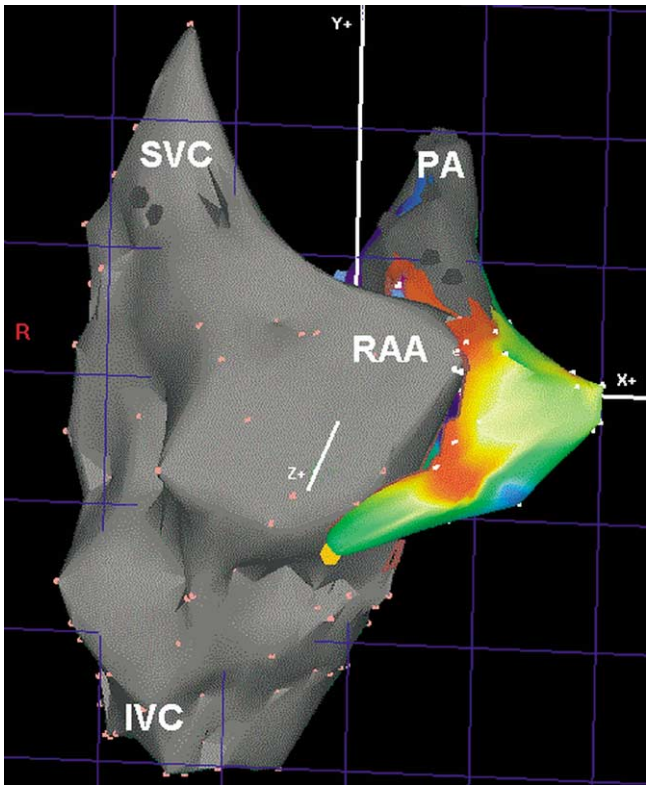

Figure 2. Carto mapping of patient 1 during atrial stimulation showing the right atrium and the pulmonary artery in gray. The ventricular infundibulum, surgically connected to the right atrial appendage, is color coded corresponding to timing of excitation, with red for the earliest excitation, followed by yellow, green, and blue. Two areas at the surgically created atrioinfundibular anastomosis are depicted in red as the ventricular insertion of the accessory pathways. *SVC*, Superior vena cava; *PA*, pulmonary artery; *RAA*, right atrial appendage; *IVC*, inferior vena cava.

mapping techniques, except in patients 1 and 4, in whom the Carto mapping system (Biosense Webster Inc, Diamond Bar, Calif) was used. In all patients, mapping of the atrium, the atrioinfundibular Fontan anastomosis, and the right infundibulum could be performed from an inferior venous access.

Accessory pathways were detected in 3 patients in a right anteroseptal or midseptal position (Figure 1). The antegrade effective refractory period was 380, 355, and 310 ms.

In patient 1 three different accessory pathways were located at the atrioinfundibular Fontan anastomosis in a superior, left lateral, and inferior position (Figure 2). In this patient an orthodromic AV re-entrant tachycardia with a cycle length of 280 and 300 ms and a self-limiting atrial flutter with an AV conduction with a mean ventricular rate of 300 beats/min could be induced (Figure 3).

In patient 4 a single accessory pathway crossing the AV Fontan anastomosis was detected. Additionally, counterclockwise atrial flutter with a cycle length of 220 ms could be induced.

All accessory pathways in our patients could be successfully treated with radiofrequency ablation (Figure 4). In patient number 1 there was a recurrence of conduction in 2 of the 3 pathways, which were successfully reablated in a second EPS 2 months later.

Additionally, in patient 4 atrial flutter was successfully treated with 2 radiofrequency ablation lines, one from the coronary sinus to the inferior caval vein and another from the coronary sinus to the surgical incision.

After these procedures, no patient showed signs of preexcitation or AV tachycardia, and antiarrhythmic drugs were discontinued. Because of frequent supraventricular extrasystoles that became overt after ablation, patient 2 is now taking sotalol. In patient 3 intermittent atrial flutter with a 2:1 AV conduction appeared, and he is taking metildigoxin and sotalol now. Patient 5 continued to be in heart failure and was treated with digoxin. Three forms of atrial flutter were recognized, but only 2 were treated successfully by means of ablation. Drug treatment with metoprolol and amiodarone could not suppress the atrial tachyarrhythmias in this patient. Three and a half years after ablation of his accessory pathway, he died from pulmonary embolism during thrombolytic therapy of a right atrial thrombus.

None of the patients demonstrated a relapse of an AV re-entrant tachycardia at follow-up up to 6 years.

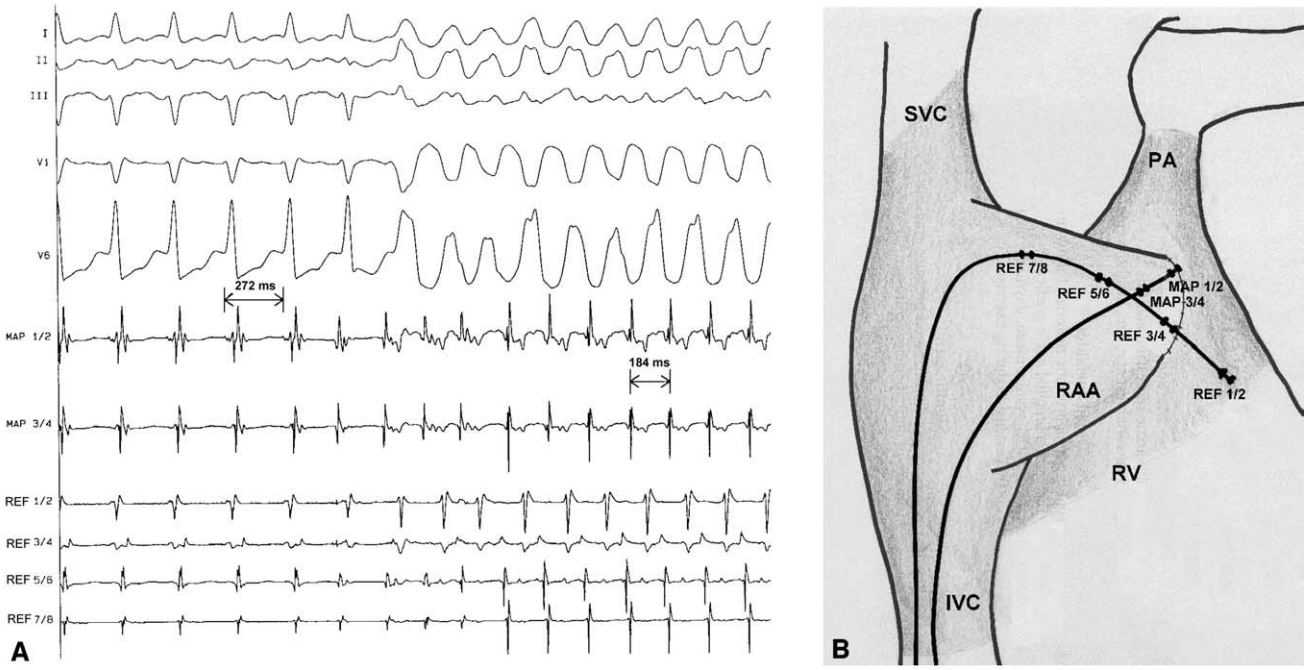

Figure 3. A, ECG of patient 1 during electrophysiologic study with I, II, III, v1, and v6 surface leads. *MAP 1/2*, Intracardiac lead near the accessory pathway showing close atrial and ventricular complexes; *MAP 3/4*, in the right atrial appendage showing only atrial complexes; *REF*, intracardiac leads across the atrioinfundibular anastomosis; *REF 1/2*, in the right ventricular infundibulum showing ventricular complexes; *REF 3/4*, at the anastomosis showing both atrial and ventricular complexes; *REF 5/6* and *REF 7/8*, in the right atrial appendage showing only atrial complexes. B, Position of the catheters. The gray area is similar to the area of the Carto mapping of Figure 2. On the left side, orthodromic AV re-entrant tachycardia with a cycle length of 272 ms is shown. During that tachycardia, ventricular impulses are conducted back to the right atrial appendage *(RAA)* through a surgically acquired pathway near the tip of the mapping catheter *(MAP 1/2)*. A single atrial premature complex induces atrial flutter and 1:1 conduction to the ventricle with a cycle length of 184 ms. Two types of ventricular complexes demonstrate either aberrancy or conduction through a second accessory pathways. *SVC*, Superior vena cava; *PA*, pulmonary artery; *RV*, right ventricle; *IVC*, inferior vena cava.

Discussion

Our study clearly shows that there is an increased incidence of accessory pathways in patients with tricuspid atresia after AV Fontan surgery, and there are 2 different types of accessory pathways: a congenital pathway as in classic WPW syndrome and a surgically created atrioinfundibular pathway.

Congenital accessory pathways have to be assumed in 3 of our patients (patients 2, 3, and 5) because of the location of the pathways. Several case reports have previously suggested an association of tricuspid atresia and WPW syndrome,[11,12] but only a few were confirmed by means of EPS (Table 3).[3,6,7,13-16] Our data confirm this association and suggest that an EPS is urgently needed in this patient group to differ between different types and locations of accessory pathways.

As in previous studies,[3,7] it is evident that all 6 congenital accessory pathways in patients with tricuspid atresia are on the right side. This confirms the theory of Misaki and coworkers[7] that the accessory pathway is on the side of the congenitally malformed AV valve, although this theory is based mainly on patients with Ebstein malformation and only on a very few number of other congenital AV valve defects.

In our patients only 1 of the 3 pathways were detected on the basis of surface ECG criteria before the Fontan operation. This is due to the well-known changes in the conducting properties in the atrial myocardium or sinus node after surgical intervention and with the high pressure and subsequent dilatation of the right atrium. Surgical injury to the AV node could slow conduction, permitting a previously occult accessory pathway to become manifest, but none of our patients showed an AV delay after ablation.

Surgically created accessory pathways were shown in patients 1 and 4 at the atrioinfundibular anastomosis. In this area, depicted in our patients by means of Carto mapping, there was no preexisting structure that can cause AV con-

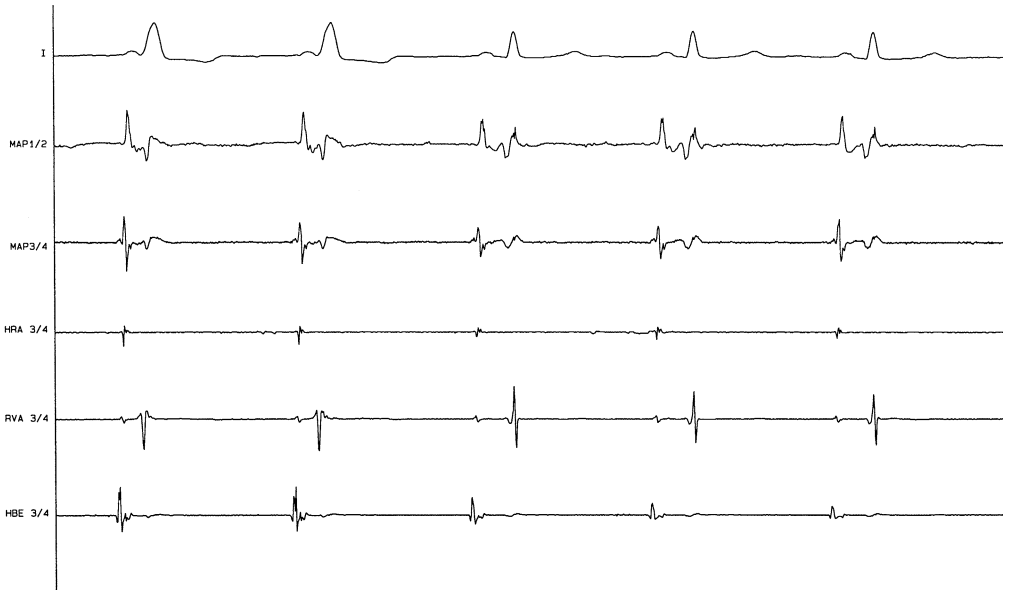

Figure 4. Intracardiac ECG of patient 3 during radiofrequency ablation. The first 2 ventricular signals show delta waves and a short PR interval of 100 ms. The following complexes depict the ECG after ablation of the accessory pathway with a narrower ventricular complex without delta wave and a normal PR interval of 160 ms. *I*, Surface lead; *MAP1/2*, tip of the mapping catheter at the accessory pathway showing signals from both the atrium and the ventricle; *MAP3/4*, more proximal lead of the mapping catheter in the right atrium; *HRA 3/4*, high right atrial lead at the right lateral wall; *RVA 3/4*, right ventricular apex lead; *HBE 3/4*, "his bundle electrode" near the AV node.

duction. These conduction pathways cross the suture line, where atrial tissue is sutured onto ventricular tissue. Conduction across a suture line was already described in an animal model,[17] in patients after heart transplantation,[18-22] and in patients after Fontan-Björk procedures.[14-16] This situation must be considered in patients after the Fontan-Björk procedure, and the surgical anastomosis must be mapped carefully when searching for an accessory pathway.

All authors reporting on patients with tricuspid atresia and WPW syndrome showed successful treatment of the accessory pathways, either by means of surgical intervention or catheter ablation.

TABLE 3. Literature review of patients with tricuspid atresia and accessory pathways confirmed by means of an electrophysiologic study

	Case no.	Morphologic diagnosis	Location AP	Treatment
Dick and coworkers, 1981[3]		TA 1b (Ebstein like)	Right posterior	OP
Misaki and coworkers, 1995[7]	1	TA 1b	Right posterior	OP
Misaki and coworkers, 1995[7]	2	TA 2b	Right anterolateral	OP
Razzouk and coworkers, 1992[14]		TA 1b	Surgical AV connection	OP
Case and coworkers, 1993[15]		TA 1b	Surgical AV connection	CA
Rosenthal and coworkers, 1997[16]		TA 1b	Surgical AV connection	CA
Drant and coworkers, 1995[6]	1-3	TA	?	CA
Hebe and coworkers, 1998[13]	1-3	TA	?	CA
This report	1	TA 1b	Lower + upper + left lateral surgical AV connection	CA
This report	2	TA 1b	Right midseptal	CA
This report	3, 5	TA 1b	Right anteroseptal	CA
This report	4	TA 1b	Surgical AV connection	CA

AP, Accessory pathway; *TA*, tricuspid atresia; *OP*, surgical intervention; *CA*, catheter ablation.

Early treatment of these accessory pathways is mandatory because atrial arrhythmias, which are often seen in the Fontan circulation, can trigger AV re-entrant tachycardia, AV re-entrant tachycardia can deteriorate into atrial flutter or fibrillation (patient 1), atrial flutter and fibrillation can be conducted 1:1 to the ventricle with the accessory pathway (patient 1), and antiarrhythmic drug treatment often fails.

Presently, the Fontan circulation is established by means of a total cavopulmonary anastomosis with an extracardiac conduit. This excludes the risk of surgically created accessory pathways but hampers easy access to the right atrium. On the basis of our data of at least 3 congenital accessory pathways (confirmed by means of EPS) in 198 patients with tricuspid atresia and older studies[11,12] without confirmation by means of EPS, there is an increased risk of accessory pathways in patients with tricuspid atresia. Therefore all these patients should be checked before the Fontan operation to exclude congenital accessory pathways. This can be performed in a minor EPS with a single ventricular lead and a single atrial-esophageal lead for pacing. In experienced hands this procedure should only slightly prolong the preoperative hemodynamic catheter laboratory investigation and should increase radiation exposure only minimally. If an accessory pathway is detected, it should be localized and ablated either in the same session or during surgical intervention, even if the patient is asymptomatic. Radiofrequency ablation is possible, even in infants with a similar success rate compared with that seen in older children.[9] Despite the higher risk of complications in infants,[23] there is consensus that ablation should be performed before surgical intervention to restrict vascular or chamber access.[24]

Those caring for patients with Fontan circulation and tachycardia must consider accessory pathways. Such patients can be treated successfully by means of catheter ablation. Patients with atrioinfundibular anastomosis have an additional risk of surgically acquired accessory pathways across the suture line.

In addition, before bypassing atrial structures with a cavopulmonary conduit, the patient should be checked for accessory pathways, and catheter ablation should be performed, even if the patient is asymptomatic.

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References

1. Gewillig M, Wyse RK, de Leval MR, Deanfield JE. Early and late arrhythmias after the Fontan operation: predisposing factors and clinical consequences. *Br Heart J.* 1992;67:72-9.
2. Porter CJ, Garson A. Incidence and management of dysrhythmias after Fontan procedure. *Herz.* 1993;18:318-27.
3. Dick MD, Behrendt DM, Byrum CJ, Sealy WC, Stern AM, Hees P, et al. Tricuspid atresia and the Wolff-Parkinson-White syndrome: evaluation methodology and successful surgical treatment of the combined disorders. *Am Heart J.* 1981;101:496-500.
4. Misaki T, Iwa T, Kuzawa Y, Murakami S, Seki M, Miyahara K. Simultaneous surgery of Wolff-Parkinson-White syndrome combined with tricuspid atresia. *Nippon Kyobu Geka Gakkai Zasshi.* 1986;34:261-6.
5. Ohtake H, Iwa T, Misaki T, Watanabe G, Mukai A, Tsubota M, et al. A successful surgical treatment of WPW syndrome associated with tricuspid atresia. *Kyobu Geka.* 1990;43:57-60.
6. Drant SE, Klitzner TS, Shannon KM, Wetzel GT, Williams RG. Guidance of radiofrequency catheter ablation by transesophageal echocardiography in children with palliated single ventricle. *Am J Cardiol.* 1995;76:1311-2.
7. Misaki T, Watanabe G, Iwa T, Yamaguchi M, Watanabe Y. Surgical treatment of atrioventricular atresia combined with Wolff-Parkinson-White syndrome. *Chest.* 1995;107:669-73.
8. Braun U, Weyland A, Bartmus D, Ruschewski W, Rath W. Anesthesiologic aspects of pregnancy and delivery in a patient following a modified Fontan procedure. *Anaesthesist.* 1996;45:545-9.
9. Blaufox AD, Felix GL, Saul JP. Radiofrequency catheter ablation in infants ≤18 months old: when is it done and how do they fare?: short-term data from the pediatric ablation registry. *Circulation.* 2001;104:2803-8.
10. Bjork VO, Olin CL, Bjarke BB, Thoren CA. Right atrial-right ventricular anastomosis for correction of tricuspid atresia. *J Thorac Cardiovasc Surg.* 1979;77:452-8.
11. Keith JD, Rowe RD, Peter V. Heart disease in infancy and childhood. New York: The Macmillan Company; 1967.
12. Schiebler GL, Adams PJ, Anderson RC. The Wolff-Parkinson-White syndrome in infants and children. *Pediatrics.* 1959;24:585-603.
13. Hebe J, Antz M, Siebels J, Volkmer M, Ouyang F, Kuck KH. High frequency current ablation of supraventricular tachyarrhythmias in congenital heart defects. *Herz.* 1998;23:231-50.
14. Razzouk AJ, Gow R, Finley J, Murphy D, Williams WG. Surgically created Wolff-Parkinson-White syndrome after Fontan operation. *Ann Thorac Surg.* 1992;54:974-7.
15. Case CL, Schaffer MS, Dhala AA, Gillette PC, Fletcher SE. Radiofrequency catheter ablation of an accessory atrioventricular connection in a Fontan patient. *Pacing Clin Electrophysiol.* 1993;16:1434-6.
16. Rosenthal E, Bostock J, Gill J. Iatrogenic atrioventricular bypass tract following a Fontan operation for tricuspid atresia. *Heart.* 1997;77:283-5.
17. Ernst RW. Pedicle grafting of the sino-auricular node to the right ventricle for treatmant of complete atrioventricular block. *J Thorac Cardiovasc Surg.* 1962;44:681-6.
18. Bexton RS, Hellestrand KJ, Cory Pearce R, Spurrell RA, English TA, Camm AJ. Unusual atrial potentials in a cardiac transplant recipient. Possible synchronization between donor and recipient atria. *J Electrocardiol.* 1983;16:313-21.
19. Anselme F, Saoudi N, Redonnet M, Letac B. Atrioatrial conduction after orthotopic heart transplantation. *J Am Coll Cardiol.* 1994;24:185-9.
20. Lai W, Kao A, Silka MJ, Halperin BD, Raitt M, Oliver R, et al. Recipient to donor conduction of atrial tachycardia following orthotopic heart transplantation. *Pacing Clin Electrophysiol.* 1998;21:1331-5.
21. Lefroy DC, Fang JC, Stevenson LW, Hartley LH, Friedman PL, Stevenson WG. Recipient-to-donor atrioatrial conduction after orthotopic heart transplantation: surface electrocardiographic features and estimated prevalence. *Am J Cardiol.* 1998;82:444-50.
22. Saoudi N, Redonnet M, Anselme F, Poty H, Cribier A. Catheter ablation of atrioatrial conduction as a cure for atrial arrhythmia after orthotopic heart transplantation. *J Am Coll Cardiol.* 1998;32:1048-55.
23. Kugler JD, Danford DA, Houston KA, Felix G. Pediatric radiofrequency catheter ablation registry success, fluoroscopy time, and complication rate for supraventricular tachycardia: comparison of early and recent eras. *J Cardiovasc Electrophysiol.* 2002;13:336-41.
24. Friedman RA, Walsh EP, Silka MJ, Calkins H, Stevenson WG, Rhodes LA, et al. NASPE Expert Consensus Conference: Radiofrequency catheter ablation in children with and without congenital heart disease. Report of the writing committee. North American Society of Pacing and Electrophysiology. *Pacing Clin Electrophysiol.* 2002;25:1000-17. Congenital and surgically acquired Wolff-Parkinson-White syndrome in patients with tricuspid atresia