

Catheter Ablation of Multiple Accessory Pathways in Duchenne Muscular Dystrophy

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A 23-year-old male with Duchenne muscular dystrophy (DMD) experienced self-limiting palpitations at age 19 years for the first time. Palpitations recurred not earlier than at age 23 years, and were attributed to narrow complex tachycardia, which could be terminated with adenosine. Since electrocardiography showed a delta-wave, Wolff-Parkinson-White (WPW) syndrome was diagnosed, ajmaline prescribed and radio-frequency catheter ablation of three accessory pathways carried out one week later. One day after ablation, however, a relapse of the supraventricular tachycardia occurred and was terminated with ajmaline. Re-entry tachycardia occurred a second time six days after ablation, and as before, it was stopped only with ajmaline. Despite administration of verapamil to prevent tachycardia, it occurred a third time four months after ablation. This case shows that cardiac involvement in DMD may manifest also as WPW-syndrome. In these patients, repeated radio-frequency catheter ablation of accessory pathways may be necessary to completely block the re-entry mechanism. (Korean Circ J 2013;43:115-118)

KEY WORDS: Arrhythmia; Tachycardia, supraventricular; Catheter ablation; Muscular dystrophies.

Introduction

Cardiac involvement is a frequent phenotypic manifestation in patients with Duchenne muscular dystrophy (DMD) and affects either the myocardium or the cardiac conduction system.¹⁾ Though cardiac rhythm abnormalities are frequently recognized in DMD-patients, Wolff-Parkinson-White (WPW)-syndrome, associated with a re-entry tachycardia, due to an accessory bypass, has only been rarely reported in dystrophinopathies.^{2,3)}

Case

In a 23-year-old male, DMD was diagnosed at age 7 years upon

Received: February 4, 2012

Revision Received: April 13, 2012

Accepted: June 26, 2012

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• The authors have no financial conflicts of interest.

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typical clinical and instrumental findings. He learned speaking at 18 months, walking at 24 months, became continent at age 3 years, and climbed stairs and became able to get up from the floor at age 4 years. Blood chemical investigations showed elevated creatine-kinase (3523 U/L, $n > 170$ U/L). Needle electromyography was myogenic and muscle biopsy showed dystrophic features and absence of dystrophin. At age 8 years tip-toe walking developed. Since age 11 years he was permanently using a wheel-chair. Since age 16 years he was no longer able to eat independently. At age 19 years he required a long-term oxygen therapy via an Oracle mouth mask, followed by a non-invasive positive pressure ventilation by means of a VS Ultra device (ST-mode, IPAP 18 mm Hg, EPAP 12 mm Hg, respiratory frequency 12, T_{min} 1.1 seconds, T_{max} 1.4 seconds, with 3 L of oxygen). Initially, he required the respirator only during the night, but later also, during the day, except when mobilized to an upright position. At age 23 years he presented with severe scoliosis and thoracic asymmetry, macroglossia, which has been only rarely described as a phenotypic feature of DMD,^{4,5)} and severe quadriplegia with residual distal motor functions, and club feet. The family history was negative for DMD, as well as WPW-syndrome.

Cardiac involvement initially manifested as palpitations at age 19 years. Electrocardiography (ECG) at that time was normal and did not show a delta-wave. At age 23 years palpitations recurred after awaking due to a narrow complex tachycardia, which could be suc-

cessfully terminated with adenosine intravenously. Blood pressure was 110/55 mm Hg. Echocardiography was normal. After resolution of the acute rhythm abnormality, ECG showed a delta-wave over all precordial recordings (Fig. 1), leading to the diagnosis of WPW-syndrome. Ajmaline was started and catheter ablation was scheduled. Ablation of three accessory pathways (right anterior-septal with anterograde and retrograde conduction, right posterior-septal with exclusively retrograde conduction, and right posterior-lateral with exclusively retrograde conduction) (Figs. 2 and 3) was successfully carried out one week after starting ajmaline without complications (Fig. 4). One day after ablation, however, a relapse of the supraventricu-

lar tachycardia occurred, which could be again relieved with ajmaline intravenously. Six days after ablation, a re-entry tachycardia recurred, which responded favorably to adenosine intravenously, but could be finally stopped only with ajmaline intravenously. On ECG, 12 days after ablation, delta-waves had disappeared, but small Q-waves over V 2-6 were recorded (Fig. 1). To prevent tachycardia, verapamil (240 mg/d) was additionally given with success. Unfortunately, the delta-wave and re-entry tachycardias recurred four months after the ablation. After switching to propafenone, neither the delta-wave nor re-entry tachycardia recurred.

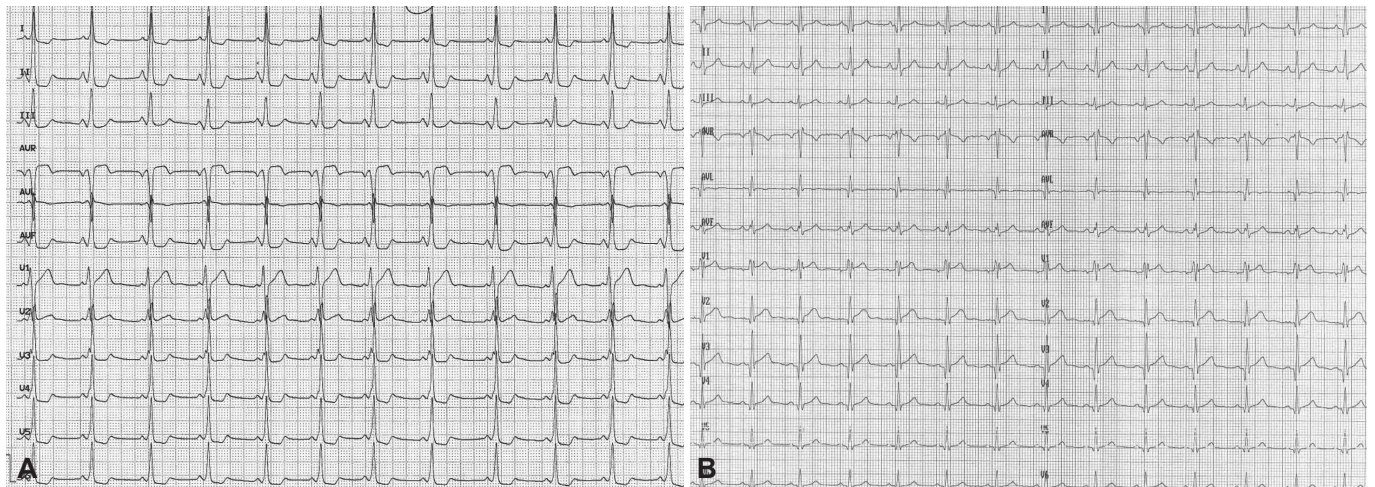


Fig. 1. ECG on admission showing shortening of the PQ-interval and a delta-wave over all chest wall recordings (A). ECG 12 days after the ablation shows incomplete right bundle branch block and small Q-waves over V 2-6, but no longer pre-excitation (B). ECG: electrocardiography.

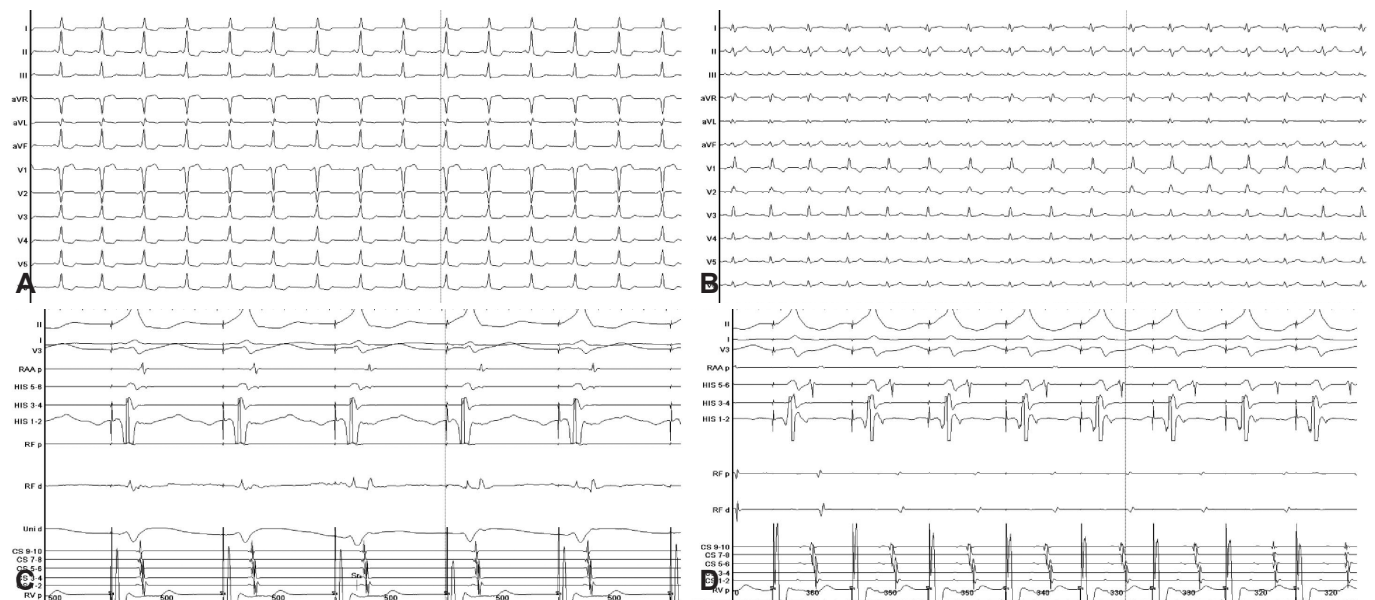


Fig. 2. Surface ECG before ablation with delta-wave (A) and after ablation without delta-wave (B). Intracardiac ECG before ablation (C) and after bump of the accessory pathway (D). The left lower panel shows change of the retrograde activation pattern during energy delivery (3rd beat from the right) with a shift to the second exclusively retrograde conducting right postero-septal accessory pathway. The right lower panel shows the retrograde activation pattern after ablation with exclusive conduction via the retrograde conducting AV-node. ECG: electrocardiography, AV: atrioventricular.



Fig. 3. The left upper panel shows change of the retrograde activation pattern during energy delivery with a shift from the right anterior septal pathway (circle) to the right postero-septal pathway (circle). The right upper panel shows the target signal for the right postero-septal pathway. The right antero-lateral accessory pathway with exclusively retrograde conduction (encircled) is shown in the left lower panel. Retrograde conduction via the AV-node after ablation is shown in the right lower panel. The ablation of the antero-lateral pathway was achieved by energy delivery via an Agilis long sheath during sinus rhythm. AV: atrioventricular.

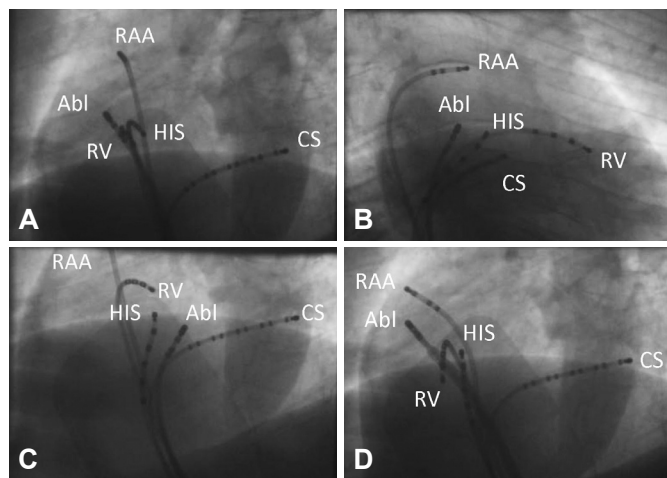


Fig. 4. Catheter position during ablation of the 1. accessory, anterograde and retrograde conducting, right antero-septal pathway in LAO 45° (A) and in RAO 30° (B), of the 2. accessory exclusively retrograde conducting right posterior-septal pathway in LAO 45° (C), and during ablation of the 3. accessory, exclusively retrograde conducting right posterior-lateral pathway in LAO 45° (D). A stable position at the tricuspid annulus was achieved only with the steerable introducer Agilis NxT (St. Jude Medical). RV: right ventricle, HIS: HIS bundle, RAA: right atrial appendage, CS: coronary sinus, Abl: ablation catheter, LAO: left anterior oblique, RAO: right anterior oblique.

Discussion

Cardiac involvement develops in the majority of the DMD-pa-

tients if they survive long enough,¹⁾ and is usually first recognized during the second decade,¹⁶⁾ although a single case developed clinical or subclinical cardiac involvement already earlier.¹⁾ Cardiac involvement in DMD usually manifests as cardiomyopathy or rhythm abnormalities. WPW-syndrome has been only rarely reported as a manifestation of cardiac involvement in DMD.²⁾ WPW-syndrome is a cardiac conduction abnormality, which is due to accessory conductive pathways between the atrium and the ventricle bypassing the atrioventricular (AV)-node.⁷⁾ ECG typically shows PR-shortening, a delta wave, and QRS-widening, and patients with WPW-syndrome develop supraventricular re-entry tachycardias or atrial fibrillation. WPW-syndrome may be associated with sudden cardiac death, of which, the risk is estimated to be 0.02%/patient/y.⁸⁾ The risk of sudden cardiac death is further increased in patients who additionally suffer from atrial fibrillation,⁹⁾ or rapidly anterogradely conducting pathways with short effective refractory period. WPW-syndrome may respond to drug therapy with adenosine⁹⁾ or ajmaline. The best and most effective and safe treatment of symptomatic WPW-syndrome, however, is ablation of the accessory pathways by radio-frequency ablation.¹⁰⁾ After ablation, high activation recovery interval-dispersion may be observed in the persisting pre-excited rhythm, but may gradually return to normal within weeks.¹¹⁾ Also, in the presented patient ablation resulted in transient resolution of palpitations and disappearance of delta-waves.

Why the described DMD-patient presented with three accessory pathways remains speculative. WPW-syndrome is widely believed to result from incomplete regression of fetal accessory atrio-ventricular pathways, a procedure which may occur in neuromuscular disorders or independently from the muscle disease. Arguments for sporadic accessory pathway formation in the presented patient are that he never complained about palpitations before age 19 years and that an ECG at age 19 years did not show a delta-wave. Possibly, the atrio-ventricular connections developed due to abnormal enlargement of cardiomyocytes, since swelling of cardiomyocytes brings atrial and ventricular cardiomyocytes together, as has been described in patients with PRKAG2 syndrome.¹²⁾ Absence of a delta-wave on the previous ECG at age 19 years could be explained with intermittent occurrence of pre-excitation. Arguments for WPW-syndrome as a cardiac manifestation of DMD are that cardiac conduction abnormalities are a frequent finding in these patients, and that with improved survival, also previously non-reported cardiac conduction abnormalities may be seen. WPW-syndrome could also be unrelated to DMD, but clinical manifestation not earlier than at age 19 years argue against such an assumption. Why reentry tachycardias recurred after ablation is most likely due to an incomplete ablation of the right anterior-septal pathway located in the proximity of the AV-node and the HIS-bundle.

This case shows that WPW-syndrome may occur in DMD-patients carrying a dystrophin mutation. The causal relation between WPW-syndrome and mutation, however, remains speculative. Repeated radio-frequency catheter ablation of accessory pathways may be necessary to completely block the re-entry mechanism in these patients.

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