Pre-excitation due to accessory sinoventricular connexions associated with coronary sinus aneurysms

A report of two cases

L M GERLIS,* M J DAVIES,‡ R BOYLE,† G WILLIAMS,† H SCOTT§

From the Departments of *Pathology and †Cardiology, Killingbeck Hospital, Leeds; the ‡Department of Histopathology, St George's Hospital, London; and the §Department of Paediatrics, West Suffolk Hospital, Bury St Edmunds, Suffolk

SUMMARY Ventricular pre-excitation occurred in two cases in which the accessory pathways between the atria and the ventricles were histologically identified as being associated with aneurysmal malformations of the coronary sinus. In one case the connexions were in the posterior wall of the coronary sinus aneurysm and were not related to the atrioventricular annulus; in the other, a connexion was situated in the anterior wall of the aneurysm in close apposition to the annulus and superficially resembled a Kent fibre. These connexions were considered to be of sinus venosus origin and to represent a modification of the muscular sheath that normally surrounds the coronary sinus but does not continue along the coronary veins. One of the posterior wall connecting bundles was composed of abnormally large Purkinje-like fibres; this may have played some role in the manifestation of the pre-excitation by reducing any mismatch impedance.

In normal circumstances the heart beat is initiated in the sinoatrial node and the impulse spreads through the atrial muscle to reach the atrioventricular node where it is delayed before passing on to the ventricular muscle through the atrioventricular bundle of His and the right and left bundle branches. This provides the only functional connexion between the inlet and outlet chambers of the heart. Pre-excitation¹ is said to occur when the ventricular response to the atrial contraction occurs earlier than would be expected if the activation had followed the usual pathway through the specialised conduction system; it is manifested by an unduly shortened PR interval on the electrocardiogram.

Various accessory pathways and connexions have been postulated or described as substrates of the various presentations of this condition (Fig. 1). Accessory atrioventricular muscle fibres bridging the fibrous tissue between atrial and ventricular myocardium may be parietal, as first described by Kent,²⁻⁷ or septal.^{8,9}

Fig. 1 Diagram showing normal and abnormal atrioventricular conduction pathways. The normal pathway is from the atrioventricular node (AVN) through the atrioventricular bundle of His (H) which penetrates the central fibrous body (CFB) and divides into the right and left bundle branches (RBB, LBB) in the ventricular septum (VS). Abnormal pathways include 1, Kent fibres; 2, Mahaim fibres; 3, James fibres; and 4, central atrioventricular pathway as described by Krikler *et al.*⁹ RA, right atrium; RV, right ventricle; LV, left ventricle.

Requests for reprints to Dr L M Gerlis, Cardiac Research Department, Killingbeck Hospital, Leeds LS14 6UQ.

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Accessory nodoventricular connexions, between parts of the atrioventricular node and the ventricular myocardium, were first described by Mahaim.¹⁰⁻¹² Bypass tracts or atriofascicular fibres passing from the atrial muscle to the atrioventricular bundle of His have been described by James,¹³ and fasciculoventricular connexions between the atrioventricular bundle and the ventricular muscle are included in the connexions described by Mahaim.¹⁰⁻¹² The preferred anatomical terminology is that proposed by the European Study Group on Pre-excitation,¹⁴ as there is considerable confusion and inaccuracy in the use of the various eponymous terms.¹⁵

When the depolarising impulse reaches the ventricular muscle other than through the atrioventricular bundle and its branches, the spread is abnormal; this is reflected in an abnormal slurring of the first part of the electrocardiographic ventricular complex, the so called delta wave. This, together with the short PR interval, is characteristic of the Wolff-Parkinson-White syndrome.¹⁶ The nature of the polarity of the delta wave in the various electrocardiographic leads enables the site of the epicardial atrioventricular connexion to be determined.¹⁷ When the atrioventricular node is bypassed, but the spread of stimulus within the ventricle is through the atrioventricular bundle and bundle branches, then the short PR interval is followed by normal ventricular electrocardiographic complexes as in the Lown-Ganong-Levine syndrome.¹⁸ Apart from the ventricular pre-excitation these various accessory pathways also provide the substrate for re-entry phenomena; these can lead to arrhythmias, which are the usual mode of clinical presentation of these syndromes. Associated anomalies have been noted, especially Ebstein's malformation of the tricuspid valve¹⁹ and hypertrophic cardiomyopathy.²⁰

We studied two cases of clinically documented ventricular pre-excitation syndrome of the Wolff-Parkinson-White type, in which there were unusual aneurysmal malformations of the coronary sinus. In both cases accessory muscular atrioventricular connexions were found within these lesions. As far as we are aware neither the coronary sinus anomaly nor this type of accessory connexion has been described previously.

Case reports

CASE 1

Clinical features

A 16 year old male dyestuffs laboratory technician presented with an eight month history of episodes of palpitation accompanied by retrosternal chest pain. The first episode lasted for about four hours. The duration of the attacks had diminished to about 30 to 45

minutes, but the frequency had increased to every three or four weeks, usually occurring after exercise. There was no relevant medical or family history; he smoked 10-15 cigarettes a day and his consumption of tea, coffee, and alcohol was moderate. Physical examination, chest radiography, and M mode and cross sectional echocardiography were all normal. Standard electrocardiography (Fig. 2) showed the appearances of a Wolff-Parkinson-White pre-excitation syndrome with a short PQ interval, and the polarity of delta waves in the various leads was in accord with anomalous atrioventricular conduction through an accessory pathway in the posterior aspect, according to the table of Gallagher *et al*¹⁷ (Fig. 3).

Treatment was started with disopyramide 100 mg and amiodarone 200 mg twice a day, and he remained well for three months when he had a further spontaneous episode of supraventricular tachycardia that required electrocardioversion. His medication was increased to disopyramide and amiodarone 200 mg each three times a day and he remained symptom free for a further six months, when he experienced another episode of supraventricular tachycardia; this responded to 5 mg verapamil intravenously. The dose of amiodarone was increased to 200 mg four times a day, and he remained well apart from two brief episodes of supraventricular tachycardia which ceased spontaneously. No episodes of atrial fibrillation or atrioventricular dissociation were noted. Graded exercise did not provoke arrhythmia.

In view of the satisfactory response to medical treatment surgery was not contemplated and electrophysiological studies were not performed. In January 1982, three years after the onset of his symptoms, he suddenly collapsed and died while walking in the street. He was 18½ years old.

Necropsy findings

Apart from the presence of a 125 g benign thymoma in the upper part of the anterior mediastinum, abnormal findings were limited to the heart. This was of normal shape, slightly enlarged (130×100×70 mm) and weighed 360 g. The apex was directed normally to the left. The pulmonary and systemic venous connexions appeared to be normal.

The atria were of solitus configuration and of equal size and normal anatomy. A 5 mm diameter 12 mm long tunnel passed upwards from the fossa ovale into the septal wall of the right atrium, but there was no communication with the left atrium. The atrioventricular valves were normal.

The right ventricle wall was slightly thickened to 8 mm and the trabeculations were well marked. The posterior papillary muscle was very small. The left ventricular myocardium and the ventricular septum were uniformly thickened to 20 mm. The free wall

Fig. 2 Case 1: (a) standard lead of electrocardiogram and (b) rhythm strip.

Fig. 3 Diagram of heart base showing position of various parietal atrioventricular accessory connexions. The asterisk indicates the position of the accessory connexion in cases 1 and 2 according to the electrocardiographic delta wave polarity. A, aorta; P, pulmonary artery; MV, mitral valve; TV, tricuspid valve; CS, coronary sinus.

was appreciably trabeculated, the papillary muscles ill defined, and many of the mitral valve chordae inserted directly into the ventricular wall.

On the posterior aspect of the heart at the base of the right ventricle immediately below the atrioven-

tricular sulcus, there was an area of cyst-like softening discernible only by palpation and without any visible indication. This occupied an ill defined area approximately 20 mm in diameter and appeared to have no tension within. Incision and subsequent complete opening showed this to be a rather more extensive lesion, extending 50 mm laterally to the right from the midline, 20 mm downwards from the atrioventricular sulcus and up to 10 mm in depth. There was partial division of the cavity by incomplete septa between the anterior and posterior walls, and the cavity was lined by a thin smooth membrane. At the upper left margin, that is approximately in the midline of the heart, this cavity communicated with the lumen of the apparently normal coronary sinus which was 4 mm in diameter and opened into the right atrium through a dilated coronary sinus orifice of 8 mm in diameter. This lesion appeared to be an abnormal (aneurysmal) posterior coronary vein (Fig. 4). The posterior wall was thin and membranous but contained extensive areas of muscle which appeared to continue into the myocardium of the wall of the right ventricle. Dissection and transillumination showed some apparent strands of muscular continuity between the posterior wall of the right atrium, the posterior wall of the coronary sinus, and the posterior wall of this ventricular venous "cyst." Dissection and transillumination of the atrioventricular rings elsewhere showed no evidence of atrioventricular muscle continuity when viewed macroscopically or with a $\times 5$ dissecting lens.

The great arteries arose normally and the pulmonary artery and aorta were each 20 mm in diameter

Fig. 4 Case 1: morphological appearance of the posterior aspect of the heart. (a) The visceral pericardium has been stripped from an area approximately equal to the extent of the coronary sinus aneurysm; the dotted area indicates the block of tissue which was taken for histological sections. (b) The posterior wall of the aneurysm has been incised and reflected upwards showing the smooth partially septated inner aspect. The opening into the right atrium is indicated by the open arrow; openings of the coronary veins are indicated by the small arrows. CS, coronary sinus; RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle; IVC, inferior vena cava; AVS, atrioventricular sulcus.

and had normal valves. The coronary arteries were normal in distribution and structure.

Histology

Serial sections showed the atrioventricular node, the atrioventricular bundle (of His), and the left bundle branch fascicles to be normal. The right bundle branch was slightly abnormal in that its origin and course were situated entirely intramuscularly, and there were early tenuous connexions between it and the working myocardium of the upper part of the

atrioventricular septum. A block of tissue was removed from the posterior wall of the coronary sinus aneurysm, as indicated in Fig. 4, extending from the left atrium to the left ventricle. Serial histological sections showed three bands of direct atrioventricular muscular connexion, two of which are illustrated in Figs. 5 and 6. One of these accessory connexions was of special interest as it was composed of very large fibres up to $50\ \mu$ in diameter, almost four times the width of the adjacent normal myocardial fibres ($10\text{--}14\ \mu$). These fibres also showed increased spacing of

Fig. 5 Case 1: serial histological sections from the block indicated in Fig. 4a. (a) The muscle fibres of the coronary sinus (CS) and the ventricle (V) are separated by a fibrous zone (F). (b) The same area as (a) at an interval of 40 μ showing the bridging continuity of a muscle strand. (Masson's trichrome stain $\times 20$.)

the myofibrils and some clear vacuolation, and they were surrounded and separated by conspicuous fibrosis. The appearances were somewhat similar to those of Purkinje fibres (Fig. 7).

CASE 2

A young girl had had a normal history apart from recurrent urinary infections associated with slight ureteric reflux since the age of 7 years. When she was 10 years old she fainted after running uphill but remained well otherwise. Six months later, however, while running at play, she suddenly collapsed, stopped breathing, and went into a tonic convulsion. She was given mouth to mouth artificial respiration during transport to hospital, where on arrival she was pulseless and without an audible apex beat. Respirations were gasping and there had been some inhalation of vomit and she was immediately intubated. An elec-

Fig. 6 Case 1: serial histological sections at 10 μ intervals ((a)–(c)) showing progressive bridging of the fibrous gap by coalescence of two portions of a prominent and discrete muscle band (arrowed). (Masson's trichrome stain $\times 20$.) CS, coronary sinus; F, fibrous zone; V, ventricle.

trocardiogram showed ventricular fibrillation; this reverted to sinus rhythm after electrocardioversion and an initial sinus bradycardia responded to atropine. She remained in sinus rhythm with a good cardiac output and made a full clinical recovery within 24 hours.

Over the next year she experienced several episodes of feeling faint, associated with pallor and palpitation, always after exercise. Several attempts were made to reproduce an attack with exercise in the hospital outpatient clinic but without success. The resting electrocardiogram did, however, show the classical appearances associated with the Wolff-Parkinson-White pre-excitation syndrome with a posterior accessory atrioventricular connexion (Fig. 8).

Almost a year after her initial cardiac arrest she again felt faint while playing in the garden, and this was accompanied by central chest pain. She was

Fig. 7 Histological sections: (a) high power view of the bridging band (arrows) shown in Fig. 6b showing the contrast between it and the adjacent working muscle. ($\times 80$.) (b) detailed higher power view of part of the anomalous band (arrows) shown in Fig. 6a showing the greatly enlarged muscular elements separated by fibrous stroma. ($\times 200$.)

Fig. 8 Case 2: standard lead electrocardiogram. (Note leads V3 and V4 are recorded at half sensitivity.)

immediately taken to hospital and on arrival she was pale and restless, and her pulse was noted to be slow and irregular. While being examined she had a cardiac

arrest, again associated with a tonic convulsion. The electrocardiogram again showed ventricular fibrillation; this did not respond to electrocardioversion, she proceeded to asystole, and all attempts at resuscitation failed. She was 11 years old when she died.

Necropsy findings

No major abnormalities were noted other than in the heart. The heart was not enlarged, and the cavities, valves, and great vessels were normal. An incision across the posterior wall of the left atrioventricular ring showed a thin walled aneurysmal sac 1 cm across which communicated with the coronary sinus through a narrow slit (Fig. 9). Lateral to this sac the sinus was absent, and the posterior cardiac vein was enlarged and extended towards the apex of the ventricle.

Histological sections traced a very large muscle connexion which slanted diagonally on the posterior wall of the heart, from the posterior wall of the left atrium to the left ventricle across the aneurysmal portion of the coronary sinus (Fig. 10).

Discussion

Accessory atrioventricular connexions and bypass tracts, of the types previously described, are common vestigial remnants of embryonic structures, but only a small proportion of these cause ventricular pre-excitation detectable by electrocardiography and very few cause symptomatic disease.¹⁹⁻²¹ It is not generally realised that apart from atrioventricular connexions, normal or anomalous, of specialised or non-specialised myocardium, there is another potential

Fig. 9 Case 2: morphological appearance of the posterior aspect of the atrioventricular junction sectioned in the sagittal plane showing the coronary sinus aneurysm (asterisk) and the slit like orifice into the coronary sinus (CS). LA, left atrium; LV, left ventricle; A, aorta.

Fig. 10 Case 2: (a) a longitudinal histological section through the coronary sinus (CS) aneurysm and the adjacent atrioventricular junction. (Masson's trichrome stain $\times 4$.) (b) Microscopical appearance of the atrioventricular junction in an area adjacent to that in (a) showing the accessory connexion (arrows). (Masson's trichrome stain $\times 20$.) AVA, atrioventricular annulus; MV, mitral valve; LA, left atrium; LV, left ventricle. *, aneurysm cavity.

avenue of communication between the proximal cardiac chamber and the ventricles and that is through the musculature of the sinus venosus. In many vertebrates the sinus venosus is a distinct contractile chamber. In mammals this separate identity is lost during fetal development, and the sinus venosus becomes absorbed into the right atrium and its muscle becomes continuous with that of the atrium. The right extension of the primitive sinus venosus, or cuvierian duct, becomes the cardiac end of the superior vena cava, and the left extension becomes the coronary sinus (Fig. 11). The proximal portion of the coronary sinus is surrounded by a spiral sheath of myocardium which is continuous with that of the morphological right atrium and is clearly a remnant of the sinus venosus musculature (Fig. 12). This muscle sheath normally stops abruptly at or shortly beyond the orifices of the entering coronary veins, but as these veins progress into intimate anatomical relation with the ventricles it remains possible that extensions of this sinus venosus muscle could be responsible for a physiological connexion. Such a circumstance might well be more likely to occur in association with a malformation of the coronary sinus. This appears to have

Fig. 11 Diagrams showing the development of the coronary sinus: (a) schematic early embryonic stage and (b) fully developed condition, as viewed from behind. The sinus venosus component is coarsely stippled. A, atrium; LA, left atrium; RA, right atrium; V, ventricle; LV, left ventricle; RV, right ventricle; SV, sinus venosus; AVA, atrioventricular annulus; SVC, superior vena cava; IVC, inferior vena cava; AzV, azygos vein; CS, coronary sinus; ACV, anterior cardinal vein; PCV, posterior cardinal vein. Asterisks indicate the positions of the sinoatrial and atrioventricular nodes.

Fig. 12 Histological sections (a) longitudinally through a normal coronary sinus and adjacent structures and (b) through the origin of a coronary vein from the coronary sinus showing the level of discontinuity of the coronary sinus muscle coat (arrow). (Masson's trichrome stain $\times 5$ (a) and $\times 20$ (b).) CS, coronary sinus; CSM, coronary sinus muscle; AM, atrial muscle; VM, ventricular muscle; AVA, atrioventricular annulus; LA, left atrium; CV, coronary vein; CA, coronary artery.

been the situation in the two cases which we have studied. In the first case the muscular connexion was located in the posterior aspect of the aneurysm and was not related to the atrioventricular annulus. In the second case the connexion was on the anterior wall of the aneurysm and was in close apposition to the fibrous atrioventricular annulus and presented a histological picture very similar to that of the more usual atrioventricular connexion or bundle of Kent; nevertheless, we consider that the underlying sinoventricular nature was common to both cases (Fig. 13).

The presence of other accessory connexions was not excluded; this would have required serial histological sections throughout the whole of the atrioventricular junction which would have been a formidable task. Dissection with transillumination in the patient in case 1 excluded major connexions, but fibres of microscopic size would not have been identified. Nevertheless, as the electrocardiographic delta wave polarity patterns indicated that the accessory conduction pathways were precisely at the position of the coronary sinus aneurysms, we consider that there is

little doubt that the connexions that we identified were responsible for the clinical features in both cases.

The large size of the fibres of one of the accessory connexion bundles in the patient in case 1 is of special interest as a possible contributory factor in the evolution of the clinical abnormality from a pre-existing symptomless anatomical malformation. Accessory tracts are not necessarily used, and Krikler and Rowland have pointed out that narrow pathways may be ineffective in stimulating a ventricular response because of mismatch impedance, in which the "cable capacity" is inadequate to depolarise the large mass of myocardium into which it is inserted.²² Enlargement of the tracts by muscle disorders such as hypertrophic cardiomyopathy may be a possible cause of activation of such latent pathways by cancelling this mismatch impedance.²³ It is not possible to know whether the Purkinje-type structure of the tract in question had always been so or whether this was an adaptive development in response to the abnormal conduction activity in which it was involved, but in either case such enlargement would be likely to enhance both the conduction rate and the cable capacity. Such changes

Fig. 13 Diagrams showing the relation of the coronary sinus muscle to the atrial and ventricular muscle (a) in a normal case, (b) in case 1, and (c) in case 2. The asterisks indicate the coronary sinus aneurysms and the arrows the accessory pathways. AVA, atrioventricular annulus; CS, coronary sinus; CV, coronary vein; F, fibrous discontinuity; RA, right atrium; RV, right ventricle; TV, tricuspid valve.

have not been described in other accessory pathways, and it is possible that they may be related to the unusual anatomical location.

Although the combination of coronary sinus aneurysm and ventricular pre-excitation is a rare condition, the finding of two almost identical examples within a short period suggests that it is worth bearing in mind especially when cases that are being considered for surgical intervention are investigated, and that during detailed electrophysiological studies placement of the coronary sinus electrode lead could quite conveniently be combined with coronary sinus contrast angiography.

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