REVIEW

When and how to assess an asymptomatic ventricular pre-excitation syndrome?

Quand et comment évaluer un syndrome de préexcitation ventriculaire asymptomatique ?

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Summary
Sudden cardiac death may be the first event in the history of Wolff-Parkinson White syndrome: this is a very rare event although as ablation of the accessory pathway avoids this risk a potentially malignant form of the disease needs to be detected. Electrophysiological studies are the most reliable method. These may be performed by a trans-oesophageal or endocavity approach from the age of six to seven years onwards. Whilst it is rare to detect a potentially malignant form the results of these studies more often enable the person to play sports or continue their job without offering radio-frequency ablation. The former however is indicated when tachycardia is induced in children over 12 years old and in adults.

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Résumé
La mort subite peut être le premier événement dans l’histoire du syndrome de Wolff-Parkinson White; il s’agit d’un événement très rare, mais l’ablation du faisceau accessoire permettant de prévenir ce risque, la detection d’une forme potentiellement maligne de l’affection s’impose. L’étude électrophysiologique est le moyen le plus fiable. Elle peut être réalisée par voie trans-œsophagienne ou endocavitaire à partir de six à sept ans. Si la detection d’une forme potentiellement maligne est rare, plus généralement les résultats de l’étude conduisent à permettre au sujet de faire du sport ou de continuer son métier sans qu’il ne soit nécessaire de proposer une ablation par radiofréquence. Celle-ci est en revanche indiquée, lorsqu’un tachycardie est induite chez l’enfant de plus de 12 ans et chez l’adulte.

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Should we be concerned about a fortuitous discovery of Wolff-Parkinson-White (WPW) syndrome in an asymptomatic person? This may be an entirely benign abnormality although sudden death has been reported as the presenting feature in these people. Several authors have shown that sudden death can be the first event in the history of the syndrome in 25 to 56% of cases reported [1–4]. The risk is nevertheless extremely low, with an estimated incidence of between 0.0% and 0.6% per year [5–8], although as ablation of the accessory pathway avoids this risk, a potentially malignant form of the disease needs to be detected. However we also know that some ventricular pre-excitation syndromes disappear spontaneously [9]. This natural progression has been shown in children with a long accessory pathway refractory period. For this reason the management of these people remains controversial.

The cause of sudden death due to WPW is understood and known to be due to the development of atrial fibrillation conducted rapidly in an accessory pathway with a short refractory period, degenerating into ventricular fibrillation (Fig. 1). The risk factors have been reported by Wellens [10]. Predisposing factors are male sex in 85% of cases, presence of circumstances precipitating high adrenergic tone such as sport and also other circumstances such as the period immediately after major surgery, and finally, electrophysiological criteria, the actual anterograde refractory period of the short accessory pathway and atrial fibrillation conducted by this bundle of Kent with cycles of less than 200 ms. Other predisposing factors have been reported although they are far more controversial: the presence of multiple accessory pathways, location of the accessory pathway in the septum, age which generally appears to be under 35 years old, cardiac glycosides and the presence of re-entry tachycardias.

The first problem is to determine whether all asymptomatic people should be investigated for a possible risk of sudden death

The only clear indication for investigation which emerges from the risk factors is people exposed to high adrenergic tone, i.e. those who take part in competitive sports or hobbies and those with an intensely physical active occupation or high stress occupations such as policemen, military personnel, firemen and any type of public transport drivers (train, plane).

Many children, adolescents and young adults play high level sports. Recent recommendations from the European Society of Cardiology require a full assessment including electrophysiological studies in these asymptomatic people before permitting them to take part in sports competitions [11]. As a result of this compulsory assessment it has become extremely rare for a ventricular pre-excitation syndrome to cause sudden death in an athlete [12]. The current difficulty is that high level sports are played in increasingly wide age ranges from young children to people over 60 years old. The problem therefore is to determine whether WPW should be investigated in all ages in life.

We know firstly that in young children atrial fibrillation is very rare under the age of 10 years old in the absence of coexisting heart disease [9,13]. Sudden deaths have been reported in 10-year old children in Pappone et al. [14] and Pagis et al. [15] series. In our own experience the two asymptomatic children who died during sports, a leisure activity in one case, were 12 years old [16]. It should be noted however that Sarrubi et al. [17] reported a case of sudden death
in an 8-year-old child although the conditions and clinical details of this child are unknown. The findings that WPW is usually benign under the age of 10 have been cast into doubt by the study by Papone et al. [14] in five to 12-year-old children which reported an apparently high incidence of induced tachycardia (60/165) and spontaneous tachycardia in these children. One case of sudden death and two cases of ventricular fibrillation were reported in untreated children, the younger of whom was 10 years old. However, it must be noted that some of these children were not truly asymptomatic as the 24 h Holter ECG records showed episodes of rapid conduction atrial fibrillation characteristic of a malignant form of the disorder and requiring treatment. The children however had no symptoms associated with these episodes of tachycardia. This demonstrates that the approach in young children in whom pre-excitation has been identified is difficult as some of them are undoubtedly not truly symptomatic and a minimum of non-invasive investigations must clearly be performed to confirm that the child has no spontaneous arrhythmias.

In adults it is conventional to recommend investigations in asymptomatic people up to the age of 35 years old [18,19]. The refractory period of the bundle of Kent tends to increase with age [20] although the risk of atrial fibrillation also increases in parallel and it is not unusual for rapid conduction syncopal atrial fibrillation to be the first manifestation in a person over 60 years old [21]. The electrophysiological findings in these apparently asymptomatic people are not significantly different from those in younger subjects [22]. Furthermore, after the age of 60, there are many situations in which adrenergic tone is raised alongside sports activities: immediate postoperative period following major surgery, accidents, serious family events or sometimes simply the Sunday afternoon dance.

It is therefore widely recommended that a pre-excitation syndrome be assessed from the age of seven years old onwards with no upper age limit depending on the person’s activities.

The second problem is to determine how to assess the prognosis of the WPW syndrome

These asymptomatic people are at low risk of serious events. We therefore must use techniques which also carry no risk but which offer good diagnostic value.

The non-invasive methods however unfortunately offer poor diagnostic value.

The surface electrocardiogram suggests a benign form of the condition if the pre-excitation is intermittent, although this is not a specific finding [10]. Features suggesting the presence of two accessory pathways call for investigation for a more serious form of the disorder.

The 24-h ECG Holter record can show tachycardia although sinus tachycardia commonly occurs in children and is difficult to interpret. The method does provide an assurance that no dysrhythmia not felt by a young child is recorded.

The exercise test is certainly the most reliable way of identifying a benign form of the disorder if the pre-excitation disappears suddenly [23]. We must be wary of progressive narrowing of the QRS complex which may incorrectly suggest that the pre-excitation has disappeared and which may lead to the diagnosis of a false positive benign WPW and miss a risk of sudden death. This situation has already been published by Attoy et al. [24] and Daubert et al.[25].

The problem with the exercise test is that the ventricular pre-excitation syndrome often does not disappear.

Disappearance of ventricular pre-excitation syndrome appearances as a result of injection of a class 1 anti-arrhythmic has been reported to suggest the presence of an accessory pathway with a long refractory period [26]. This test has now been abandoned as it unfortunately offers low diagnostic value and has risks in the presence of underlying heart disease.

Electrophysiological studies are now the reference investigation to assess the prognosis of the ventricular pre-excitation syndrome. The investigation has high diagnostic value to identify at risk people [18] and is the most sensitive and specific method. Induction of atrial fibrillation conducted rapidly by the accessory pathway (over 240/min at baseline base state and over 300/min on isoprenaline) [10] can identify a person with a malignant form of the disorder. Papone et al. [18] recently added triggering of re-entry tachycardia as a risk factor for events although this was above all a risk factor for developing a dysrhythmia. It should be noted that if the person is truly asymptomatic, orthodromic tachycardia is very rarely induced, in 10 to 25% of cases [16,27]. On the other hand, atrial fibrillation is induced as commonly in asymptomatic as in symptomatic people and is found in 20 to 50% of cases depending on the method used. Atrial fibrillation is more readily induced by conventional intracavity electrophysiological studies than by the trans-oesophageal approach [28]. The incidence of forms classified as malignant ranges from 15 to 25% of cases depending on the studies and electrophysiological study protocol used. This also means that 75 to 80% of the people have a benign form of the disorder which must be acknowledged and in whom sports can be permitted without the need for ablation.

Electrophysiological studies were however controversial until 2003 as until that time the identifying malignant forms of the disorder had not been found to be associated with a true risk of event in patient follow up [29,30]. It was Papone et al. [18] who demonstrated in a large series of 224 adolescents and young adults that 3 patients with rapid induced atrial fibrillation died because they had ablation of their accessory pathway. Papone et al. also reported the same high risk in 5 to 12-year-old children [14].

Electrophysiological studies themselves have a few problems. General anaesthesia may be required in young children and the complications of catheterization are not particularly rare in children [31] or in adults, being found in 3% of cases in the Italian study [14] in children under 12 years old. The endocavity approach requires harmful irradiation in an asymptomatic child [32]. The risk of inducing atrial fibrillation is over-estimated in children [14]. For this reason depending on the electrophysiologists experience the trans-oesophageal approach may be preferable in asymptomatic people as it produces exactly the same information [33,34] and may be performed with the same stimulation mate-
rial and same electrophysiology suite as for the endocavity approach: above all it can be performed without hospitalisation, is fast, carries no risk of irradiation and can be performed in children from the age of 6 and over [13]. If a malignant form is found, the person can be recalled secondarily for radio-frequency ablation of the accessory pathway although the likelihood of this is small. The technique also has disadvantages: it can cause chest pain although this is now far less as stimulation can be performed with a biphasic pacer: it is recommended that parents are present with young children; as the most difficult problem is making the child swallow the oesophageal probe. In approximately 1 to 5% of cases it may not be possible to stimulate the atrium from the oesophagus as the oesophagus is known to lie variably close to the left atrial wall. Finally, any induction of atrial fibrillation through this approach limits the assessment of the shortest cycle conducted by the accessory pathway in atrial fibrillation, which until 2003 was considered to be the reference information to assess the risk from the disorder.

The question of follow up and whether or not there is a need to repeat the investigation arises in children: the investigation should only be repeated very occasionally, and only if symptoms develop later and possibly in children under 12 years old in whom orthodromic tachycardia has been induced as in the case of a long refractory period in a child up to the age of 12 the accessory pathway may degenerate spontaneously [9]; this is seen above all in children under one year old, in 50% of cases although is only possible beyond the age of 12 years old if the accessory pathway has a long refractory period: if the refractory period is short, electrophysiological findings change little over time.

Conclusion

As they are easy to perform, electrophysiological studies, which are the most reliable means of detecting a malignant form of pre-excitation, are widely indicated at all ages. It is generally rare to find a potentially malignant form and the electrophysiological studies in general enable the patient to be permitted to play sport and to continue their job without offering radiofrequency ablation. This is of course indicated if a tachycardia is induced in children over 12 years old.

References

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