Undetected cardiac lesions cause unexpected sudden cardiac death during occasional sport activity

A report on 80 cases

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The retrospective analysis of 1500 forensic autopsies after sudden cardiac death showed that 80 (77 men, three women) had died following sport, for which they had been inadequately trained. The chosen sport (both dynamic and static), and the cardiac pathology discovered during autopsy make it possible to divide the population into two groups. Group 1 were those under 30 years of age (27 cases) engaged in jogging, gymnastics, rugby, tennis and boxing who suffered from hypertrophic cardiomyopathy (29.6%), arrhythmogenic right ventricular cardiomyopathy (25.9%), non-atherosclerotic (14.8%), aortic stenosis (7.4%), atrial septal defect (3.7%), stenosing coronary atherosclerosis (3.7%), and structural abnormalities of the His bundle (3.7%). Group 2 were those over 30 years of age (53 cases), engaged in swimming, cycling, jogging and football. The cardiac lesions responsible were stenosing atherosclerotic coronary disease (49%), non-atherosclerotic coronary disease (1.8%), hypertrophic cardiomyopathy (20%), obstructive cardiomyopathy (4.8%), structural abnormalities of the His bundle (7.4%), myocardic bruise scar (4%), and arrhythmogenic right ventricular cardiomyopathy (3.7%). In both groups, dilated cardiomyopathy occurred with identical frequency (11%).

Conclusions The lesions discovered are the same as those identified in professional athletes, when the body tries to avoid mortal rhythmic decompensation in the case of an over-loading volume and tension during an ill-adapted effort. Forensic autopsy should establish these anomalies because the transmissible genetic characteristics of some of them could underline the need for check-ups in other members of the family.

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Key Words: Sport, sudden cardiac death.

Introduction

Sport at school is becoming more and more common and membership of sport associations is increasing every year. In spite of this trend, between 1200[1] and 1500[2] cardiovascular deaths in subjects engaged in sport are recorded each year in France. Two forms of sudden death are included in these figures: expected death in individuals who have previously presented cardiovascular symptoms, and the unexpected sudden death of individuals who were previously considered healthy. The rare post-mortem data available (300 cases to date) do not make it possible to evaluate precisely the relative prevalence of these two situations. The distinction is, however, of the utmost importance. Indeed, a precise clinical examination, guided by a detailed interrogation, in which modern exploratory techniques are applied, would reveal the majority of cases of occult cardiovascular disease, and thereby prevent a large number of unexpected sudden deaths and the concomitant serious psychological damage caused to the deceased’s friends and family. What would the cost be of implementing such a scheme, and could this cost be borne by the community as a whole[3]?

Material and methods

Between January 1980 and December 1995, 1500 heart-lung block subjects, aged from 2 to 65 years, were examined by order of Public Prosecutors, following unexpected sudden death (defined as less than 1 h) between onset of symptoms and death occurring in subjects with no particular history and excluding traumatic and toxic causes. Eighty of these subjects died during recreational sport. Autopsies, performed at the forensic medicine institute, revealed no cerebral or
abdominal injuries or acute intoxication potentially causing death. The heart–lung blocks underwent general examination, followed by histopathological analysis, in accordance with a precise protocol[4,5].

The population studied included 80 sportspersons, 77 males and three females, with an average age of 35·8 ± 14·6 years. All cases had appeared completely asymptomatic (according to police reports and families) prior to the accident. The subjects took part in relatively vigorous sport (running, gymnastics, tennis, boxing, swimming, cycling, jogging, soccer and rugby). The frequency of physical activity was variable. Among the under 30s (group 1) 27 subjects practised sport sporadically either in or out of the educational context. Among the over 30s (group 2) 53 practised sport at least once a week and the other seven, two or three times a month.

### Results

For all subjects, the weight of the heart was found to be considerably greater than average for the same age group, at 343 ± 104 g for the under 30s (normal mean weight is 200 g for the 10–20s and 300 g for the 20–30s) and 525 ± 120 g in the over 30s (normal mean weight is 350 ± 30 g). The lesions discovered were classified as coronary lesions (details are given in Table 1), myocardial lesions (Table 2) and rare lesions. The latter include:

- An aortic rupture in a 28-year-old subject presenting with Marfan’s disease.
- An inter-atrial communication associated with mitral valve prolapse and fibrosis of the His bundle trunk in an adolescent male who died while playing soccer.
- Two cases of congenital aortic bicuspid stenosis: one in a 14-year-old boy who died during a cross country run, and the other in a 16-year-old boy, who died during a bicycle race.

### Discussion

The 80 observations collected over a 15-year period represent an average of 5·3 incidents per year in the

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**Table 1  Coronary lesions**

<table>
<thead>
<tr>
<th>Coronary lesions</th>
<th>G1</th>
<th>G2</th>
<th>With myocardial infarct</th>
<th>Without infarct</th>
<th>Associated with</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Acute</td>
<td>Old fibrous scar</td>
<td>Acute and old fibrous scar</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Non-atherosclerotic</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Muscle bridging in LAD</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aneurysm (Kawasaki’s disease)</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Congenital abnormality*</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Atherosclerotic</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stenosis of 3 trunks</td>
<td>1**</td>
<td>13</td>
<td>1</td>
<td>4</td>
<td>7</td>
</tr>
<tr>
<td>Stenosis of 3 trunks + thrombosis in 1 trunk</td>
<td>4</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Stenosis of 2 trunks</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stenosis 1 trunk (LAD)</td>
<td>6</td>
<td>1</td>
<td></td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>Stenosis + thrombosis in 1 trunk</td>
<td>2</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>3</td>
<td>27</td>
<td>3</td>
<td>6</td>
<td>2</td>
</tr>
</tbody>
</table>

*The left coronary artery arising from a common ostium with the right coronary artery. **Subsequent enquiry revealed a family history of hypercholesterolaemia.

FHB=fibrosis of the His bundle; MVP=mitral valve prolapse; LAD=left anterior descending coronary artery.

**Table 2  Myocardial lesions**

<table>
<thead>
<tr>
<th>Main lesions</th>
<th>Isolated G1</th>
<th>G2</th>
<th>Associated with</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>His bundle fibrosis</td>
<td>Intra-mural vessel anomalies</td>
<td>Mitral valve prolapse</td>
</tr>
<tr>
<td>Hypertrophic cardiomyopathy</td>
<td>2</td>
<td>5</td>
<td>6</td>
</tr>
<tr>
<td>Dilated cardiomyopathy</td>
<td>3</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Asymmetric hypertrophic cardiomyopathy</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>His bundle structural anomalies</td>
<td>2</td>
<td></td>
<td>3</td>
</tr>
<tr>
<td>Tawarien mesothellosma</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ARVC</td>
<td>3</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Scarred myocardial bruising</td>
<td>2</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>6</td>
<td>13</td>
<td>12</td>
</tr>
</tbody>
</table>

LAD=left anterior descending coronary artery; ARVC=arrhythmogenic right ventricular cardiomyopathy.
Lyon and Saint-Etienne regions, with a population of two million inhabitants. These observations represent 5.3% of the cardiac sudden deaths in the region for this period. Similar proportions have been reported by Kennedy in St Louis[6] and Burke et al.[7] in Maryland, U.S.A. The predominance of men in the series concords with previously reported data[8–9]. The fewer risk factors and the higher proportion of women engaged in non-physical leisure activities might explain this difference.

The majority of the pathological situations observed (68 cases) were covert, and only detectable at autopsy. In 12 cases (coronary injuries with previous necrosis and congenital aortic stenosis), the injuries were either undetected or were considered to be not serious enough to interfere with the practice of sport.

Such cardiac injuries can remain asymptomatic throughout a lifetime. Brutal rhythmic breakdown (tachycardia followed by ventricular fibrillation in the majority of the cases, asystoles being rare) can also occur unexpectedly during volumetric or pressure overload during effort and is favoured by dehydration (with or without hypokalaemia) and by treatment with medications such as tricyclics and antidepressants[8–14].

The predominant role of non-atherosclerotic coronary artery disease in cardiac sudden death in the under 15s (15% in our series) is indicated in nearly all the series reported in literature[6,11]. In two of the cases of atherosclerotic coronary heart disease (one in the under 30s and one in the over 30s), a family history of hypercholesterolaemia was revealed, leading to examination of other members of the family who have thus been able to benefit from appropriate treatment. Coronary atherosclerosis, at 52%, is the most frequently observed pathology in the over 30s, but is lower than the 80% of sudden deaths during effort attributed to this cause in the U.S.A[6]. This difference may be due to our selection criteria, which excluded known cardiac cases from the study, and possibly ‘the French paradox’.

**Myocardial injuries**

Hypertrophic concentric cardiomyopathy is, in agreement with published findings[10,11], the most frequently observed condition reaching 29% in the under 30s and 20% in the over 30s. Only one case of obstructive cardiomyopathy (asymmetric hypertrophy) was found in our series. This aetiology is usually observed more frequently[12]. The absence of reports of dilated cardiomyopathy in the literature is surprising as nine cases were observed in our series (three in the under 30s and six in the over 30s).

Arrhythmogenic right ventricular cardiomyopathy (Uhl’s partial anomaly)[13] is the second most frequently observed condition, with seven (24%) cases in group 1 and two cases in group 2. What is interesting about myocardial lesions is that in the majority of cases several associations result in a fragile rhythmic state. For example, hypertrophic concentric cardiomyopathy was isolated in seven cases and associated with His bundle trunk alterations and intramural arteriole injuries in 12 cases[14,15]; dilated cardiomyopathy was isolated in five cases, associated with His bundle injuries in two cases, and with mitral valve prolapse in two cases; arrhythmogenic right ventricular cardiomyopathy was isolated in two cases (involving the anterior wall of the right ventricle — hypoplasia or aplasia of the myocardium, displaced by fatty infiltrations with a few crossing fibres); the injuries were more complex in the other seven cases, associating anomalies of the right ventricular wall with left ventricular hypertrophy in three cases, and lesions of the His bundle and intramural arterioles of the upper interventricular septum in the other four cases[16,17]. Structural anomalies of the His bundle trunk were observed in seven cases. The damage was morphologically very specific[18–24]; the cells of the His trunk bundle and the contractile myocardial cells of the crestal septalae were rarified; the remaining cells were wrapped in reticular fibrosis or dislocated by blocks of mutilating fibrosis. In both regions the small arteries presented thickened walls, sclerohyaline and luminal narrowing.

Scarred myocardial bruising is a rare injury. It was observed twice in the series, once in a man of 38 who died during a game of soccer and once in a man of 50 who died during a game of rugby. Examination revealed retracted subepicardiac and transmural fibrous scarring of the posterior wall of the left ventricle, which sometimes attained the posterior-septal diedra. Microscopic examination revealed granular haemosiderotic pigmentation, indicating previous haemorrhage. The history of both cases revealed a frontal sterno-thoracic shock.

**Conclusions**

Although death during sport represents a small proportion of unexpected sudden deaths (80 out of 1500 cases in our series), it remains a major medico-legal problem. The study of these deaths leads to two conclusions. First, during autopsy, rigorous pathological examination may detect unsuspected injuries. This can help to relieve the anxiety of the friends and family of the deceased. Second, given the genetic nature of certain anomalies[25], the examination of other family members can be advised if appropriate.

Clinical examination, guided by detailed questioning, taking into account minor symptoms and using modern diagnostic methods (based on the patient’s history, baseline electrocardiogram and echocardiography) would detect the majority of cases of occult heart disease, and could thereby help to prevent the catastrophe (particularly at school during physical education classes) of sudden death. The gradual atrophy of school-based medicine over the last few decades in France must be recognised as being partially responsible for the unacceptable level of cardiac sudden death in young people, and those involved in the promotion of cardiovascular health cannot be encouraged too strongly to lobby to reverse this trend.
References