Inherited cardiomyopathies

Editor—It was interesting to read Staikou and colleagues’ excellent review of Perioperative management of hereditary arrhythmogenic syndromes. This group of disorders seems to have previously been under represented in mainstream anaesthetic literature.

The collective incidence of the group of disorders that are described means that all consultant anaesthetists will have anaesthetized a number of patients who will subsequently present with one of these disorders. The fact that presentation is often with sudden death raises obvious questions as to whether a number of these deaths could be prevented by earlier identification.

The majority of those presenting for anaesthesia with such an underlying condition are likely to arrive with no confirmed diagnosis. Diagnosis of these conditions may be difficult as a large number of genotypes can result in different phenotypic variations, which may be characterized under one diagnosis. Variable penetrance and the fact that presentation of these congenital abnormalities is often not before adolescence, or later in life, makes the identification of familial patterns difficult.

Traditional teaching has been that ventricular extrasystoles are not uncommon in middle age and in patients with no ischaemic heart disease or obvious cardiomyopathy are usually of no clinical significance. Similarly, occasional short runs of bigemini noted on ECG monitors are often assumed to be benign if they resolve spontaneously and do not result in any detectable short-term consequences.

Anaesthetists watching ECGs for extended periods of time during otherwise uneventful cases are ideally placed to identify and highlight certain individuals for investigation. Is it now time for anaesthetists to have a higher index of suspicion when they observe frequent ventricular ectopy that do not result in short-term detrimental effects? A postoperative discussion to elicit any family history of unexplained or early cardiac death, 24 h ECG monitoring, and an echocardiogram would seem to be appropriate steps to take in any patient showing abnormal ectopy. The challenge now, in an era of finite resources, must be to define the level at which to act.

Declaration of interest

None declared.

W. Fish*
Cornwall, UK
*E-mail: william.fish@rcht.cornwall.nhs.uk

Reply from the authors

Editor—We thank Dr W. Fish for his interest in our review and his constructive comments regarding the role of anaesthesiologists in early diagnosis of hereditary arrhythmogenic syndromes in suspicious cases.

We agree that arrhythmogenic syndromes are underdiagnosed, since they may be asymptomatic for a long time. Thus, several anaesthesiologists have probably provided, or will provide at some time, anaesthesia to an undiagnosed patient. Since most of the times the postoperative outcome of these patients is good, an intraoperative arrhythmia, especially if short-lived and self- or pharmacologically resolved, may not be further investigated, either because its clinical significance is underestimated by the physician, because the medical costs will increase, or both.

It is true that the anaesthesiologists can be adequately prepared for the perioperative management of patients diagnosed with an arrhythmogenic syndrome, in terms of preoperative clinical optimization of the patient, a multidisciplinary approach, and a case-tailored anaesthetic plan. On the contrary, uncontrollable difficulties may arise during management of undiagnosed patients whose syndrome is unveiled perioperatively. This explains the fact that most reported perioperative deaths have occurred in undiagnosed patients, as shown in the tables of our review.

Perioperatively, anaesthesiologists should maintain a high level of suspicion for arrhythmogenic syndromes, especially in cases with a personal or family history of arrhythmias, syncope, or sudden cardiac arrest. A detailed history is of paramount importance in revealing suspicious symptoms indicating paroxysmal arrhythmias—such as palpitations, dizzy spells, fatigue, impaired memory, syncopal episodes—especially if they occur under excessive sympathetic or parasympathetic activation, that is, physical exercise, emotional stress, or vagal manoeuvres. If an arrhythmogenic syndrome is suspected before operation, the patient should be further investigated before he/she undergoes a scheduled surgical procedure. This is highlighted by the case reported by Nakamura and colleagues regarding a patient with asymptomatic bradycardia resistant to atropine manifested just before induction of anaesthesia. The scheduled surgery was postponed and further cardiological investigation revealed sick sinus syndrome. After placement of a temporary cardiac pacemaker, the patient underwent uneventfully a 9 h neurosurgical procedure.
Of course, in many cases, neither the medical/anaesthetic history nor the preoperative examinations raise suspicions for an arrhythmogenic syndrome. A characteristic case is the one reported by Hirata and colleagues, regarding a surgical patient with undiagnosed sick sinus syndrome and normal preoperative cardiac examinations, including a Holter electrocardiogram. The syndrome was unveiled after induction of general anaesthesia and was confirmed a few months after operation by a diagnostic new Holter electrocardiogram.

In patients with unexplained, suspicious intraoperative arrhythmias, even if they resolved without further complications, postoperative 24 h haemodynamic monitoring and further cardiological investigation, although associated with increased costs, would probably be useful in revealing an arrhythmogenic syndrome. If a sudden perioperative death occurs, postmortem investigation and—if indicated—familial genetic screening should be performed. In these cases, the anaesthesiologists may also play a significant role in announcing the death, explaining, informing, and even guiding the family members towards investigations which may be lifesaving for them, if a hereditary syndrome is diagnosed and thus treated early.

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C. Staikou*
K. Chondrogiannis
A. Mani
Athens, Greece
*E-mail: c_staiou@yahoo.gr

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**FIO2 and studies on oxygenation during one-lung ventilation**

Editor—We read with interest the study by Rozé and colleagues comparing the effects of two ventilation strategies on oxygenation during one-lung ventilation (OLV).

Although not explicitly stated, the authors seem to have used variable levels of FIO2 across subjects during OLV. However, to study the effects of changes in ventilation strategy (or any other intervention) on oxygenation during OLV, it may not be advisable to vary FIO2 across subjects and present data as PAO2/FIO2. It rather may be helpful to use a constant and high FIO2 in all patients and present data as PAO2.

Why is it problematic to vary FIO2 and present data as PAO2/FIO2? This is because the relationship between PAO2/FIO2 and FIO2 is not linear and may vary considerably with FIO2. The variation would be most apparent in patients with large shunts and ventilation/perfusion abnormalities, pathologies prevalent in the thoracic surgical patient population. A low FIO2 in patients with low ventilation/perfusion ratio may, for example, increase venous admixture. Thus, using variable levels of FIO2 in a patient population with respiratory disease and different shunt fractions may generate excess variation in PAO2/FIO2 values unrelated to the intervention. The cross-over design in this study may have averted gross variation with respect to the intervention but does not rule out excess interindividual variation.

Why is it better to use not only a constant but also a high FIO2 (>0.8) and present data as PAO2? This is better because while using high FIO2 even small changes (increase or decrease) in shunt fraction, induced, for example, through the intervention under study, would predictably lead to large changes in PAO2. While using low FIO2 similar changes in shunt fraction may lead to comparatively smaller changes in PAO2 and thus less chances of obtaining statistically significant results. This can be readily appreciated by studying the iso-shunt lines, the graphic interrelationship between PAO2, shunt, and FIO2.

During clinical OLV, however, we too advocate using low FIO2 compatible with sufficient oxygenation.

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W. Karzai1*
U. Klein2
1Bad Berka, Germany
2Nordhausen, Germany
*E-mail: karzai@zentralklinik.de

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**Reply from the authors**

Editor—We thank Karzai and Klein for their interest in our article. We totally agree with them regarding the interpretation of the PAO2/FIO2 ratio. It is important to clarify that this