

ESC recommends DNA analysis in post mortems of young sudden death victims

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ESC Guidelines published today recommend DNA analysis as a fundamental component of post mortem assessment in young sudden death victims. Identification of a genetic cause helps to quickly diagnose and protect relatives.

The Guidelines are published online in *European Heart Journal* and on the ESC Website and are the European update to the 2006 European/American guidelines.3 They focus on preventing sudden <u>cardiac death</u> in <u>patients</u> with <u>ventricular arrhythmias</u>.

"For the first time these guidelines have incorporated the concept proposed by several consensus documents that DNA analysis should be a fundamental component of post mortem assessment in young sudden death victims," said Professor Silvia Priori, Chairperson of the guidelines Task Force. "The molecular analysis helps to identify the presence of genetic diseases that can occur in a structurally normal heart and therefore cannot be identified during autopsy. Identification of a genetic cause as a substrate for a sudden death facilitates the early diagnosis of affected relatives and may help protect them through a personalised approach that spans from lifestyle modifications to the early use of therapies."

Another innovation is the introduction of "emerging recommendations" which are not yet supported by evidence strong enough for a Class I recommendation but which are highly promising. One example is the Class IIa recommendation, level of evidence C, that supports the use of



the old antiarrhythmic drug flecainide in addition to beta-blockers for patients with a diagnosis of catecholaminergic polymorphic ventricular tachycardia who experience recurrent syncope or polymorphic/bidirectional VT while on beta-blockers and have risks/contraindications for an <u>implantable cardioverter defibrillator</u> (ICD).

"This new indication for an old drug is particularly relevant, as it provides an additional therapeutic option for patients with an 'orphan' disease with a limited number of available treatments," said Professor Priori.

With respect to new technologies, two Class IIb recommendations are introduced that cautiously inform the medical community about new treatment options that are still under investigation but could be used in select patients when alternatives are ineffective, unavailable or contraindicated. First, wearable cardioverter defibrillators may be considered for adult patients with poor left ventricular systolic function who are at risk of sudden arrhythmic death for a limited period and are not candidates for an ICD.4 Second, the subcutaneous ICD may be a useful alternative to a transvenous defibrillator when venous access is difficult, after the removal of a transvenous defibrillator due to infections, or in young patients with a long-term need for ICD therapy.

One of the most important recommendations is to identify patients with ischaemic heart disease who are at high risk of sudden cardiac death and may benefit from an ICD. An eight year follow-up of the MADIT II study found that patients with an ejection fraction below 30% and mild to moderate heart failure (NYHA class II and III) had a long-term improvement in survival with an ICD. ESC Guidelines published today recommend re-evaluation of left ventricular function at 6-12 weeks after myocardial infarction to assess the potential need for primary prevention ICD implantation.



"This recommendation is based on the fact that many patients with reduced ejection fraction early after a myocardial infarction will experience an improvement in their condition over time and therefore do not require the implantation of an ICD," said Professor Carina Blomström-Lundqvist, co-Chairperson. "Reassessment of ejection fraction is critical to ensure that all efforts are made to avoid unnecessary ICDs while at the same time identifying patients whose ejection fractions remain low even after medical therapy and who need an ICD."

Cardiac resynchronisation requires further study in a number of areas. "Its use in patients with atrial fibrillation has not been defined outside of observational datasets and there is a clear need for adequately powered randomised trials in this patient group," said Professor Blomström-Lundqvist. "Furthermore, whether the duration or the 'morphology' of the QRS complex is the most important predictive factor for response to therapy remains a challenging issue requiring more extensive investigations."

Professor Priori concluded: "The guidelines emphasise that early diagnosis of disorders that may predispose to sudden cardiac death can save lives. Screening in the general population may be expensive and not all experts support its use. But there is now a broad consensus that cascade screening should always be performed in families of affected individuals and in families with a history of <u>sudden cardiac death</u> in young people."

More information: 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. European Heart Journal. 2015. <u>DOI:</u> 10.1093/eurheartj/ehv316

ACC/AHA/ESC 2006 guidelines for management of patients with



ventricular arrhythmias and the prevention of sudden cardiac death. Europace. 2006 ;8:746-837. <u>DOI: 10.1093/europace/eul108</u>

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