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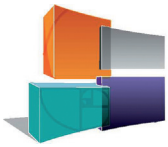

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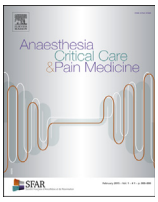

Letter to the Editor

Management of malignant hyperthermia in France: Current organisation

Introduction: Malignant Hyperthermia (MH) is a pharmacogenetic disorder of skeletal muscle that results as a hypermetabolic response to exposure to halogenated anaesthetic agents with or without depolarizing muscle relaxant (the direct role of depolarizing MR remains controversial). Its incidence ranges from 1/5000 to 1/100,000 anaesthesias. It is a specific anesthesic complication, and its management during crisis depends on anaesthesiologist's intervention. Early recognition of the signs of MH and specific treatment with Dantrium are critical to decrease morbidity and mortality of MH during anaesthesia. Intense MH crisis that is associated to symptoms like hyperthermia, muscle rigidity, hypercapnia can occur early after the beginning of the anaesthesia. The use of new halogenated anaesthetic agents, such as sevoflurane and desflurane, modified the presentation of this classical acute crisis. Indeed they allow delayed onset crisis (until two hours after the end of anaesthesia), a lesser increase of body temperature and hypercapnia, and reduced (or even prevent) muscle rigidity. MH diagnostic remains difficult in many cases and specific knowledge is necessary to avoid misdiagnosis. To help anaesthesiologists in crisis' management, specialised websites (such as www.emhg.org or sfar.org) edited recommendations and procedures. Nevertheless the only way to establish a reliable diagnosis is a genetic blood test and/or muscular biopsy that must be performed by specialised physicians. However French bioethics laws allow the communication of a genetic blood results only to the former prescribing doctor. Moreover, the absence of specified patient's phenotypes, the dominant genetic transmission of MH and the high variability in genetic penetrance, require a specific knowledge of MH. This would improve the management of the patients MH sensitive and their family members. Thus, the aim of this study was to describe the current management of patients with suspected MH in France.

This study consisted in a survey of knowledge among French specialised centres in the follow-up of patients suspected of MH or those (or their family members) with a confirmed diagnostic of MH.

Overall, four centres are specialised in post-crisis management of MH namely: Lille, Paris, Grenoble and Marseille. Lille and Marseille share the ability to perform muscle biopsy and IVCT (in vitro contracture test) Halothane-caffeine and only one centre (Grenoble) performs the genetic blood tests. We retrospectively analysed Grenoble centre data's between January 2016 and December 2017.

Results: In 2016, 162 genetic blood tests were performed, 52 for clinical MH suspicion and 110 for the family's follow up. Among the 52 propositus, 32 (61.5%) were addressed by a MH centre and 7 had a mutation recognised for MH.

In 2017, 142 genetic blood tests were performed, 53 for clinical MH suspicion and 89 for the family's follow up. Among the 53 propositus, 35(66%) were addressed by a MH centre and 18 had a mutation recognised for MH.

In about 35% of cases, the genetic blood test was prescribed by the anaesthesiologist (or by one of the members of this physician team) who was caring for the patient when the crisis occurred. In remaining cases, the test was prescribed by a physician of specialised centres. Among patients detected positive to MH by exterior physicians, about 30 % are never referred to MH specialised centres due to Bioethics French law which clearly state that results of genetic test cannot be communicated to other physician except the former who prescribed the test. Though, many patients and their family may still be inadequately monitored, especially when a specific mutation of the *RyR1* is shared by family members. Moreover, these patients could not be recruited for the contribution of France database. This might introduce some bias in studies' results interpretations obtained from this database.

More informative studies have to be lead for the management of MH in France. This includes: widespread information to of all physicians about the importance of addressing patients to a specialised centre when suspecting a MH crisis. It will also make possible the prevention of the risks for the family members to trigger a crisis (given the genetic nature of the disease) when either individual or familial histories suggest a past MH crisis.

Disclosure of interest

The authors declare that they have no competing interest.

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