1. INTRODUCTION

SUDEP is a fearful complication of epilepsy, and discussions about ways to prevent, or to cope with the psychological burden of the families, whose children suffer from a chronic convulsive disorder, are becoming more and more frequent. Several risk factors have been mentioned; moral and legal issues have been widely argued and contradictory positions are a rule rather than an exception. Now, when someone has to handle the issue with newly diagnosed epilepsy, of course the medical position would be different from a situation when the disorder is old-dated. The truth-telling philosophy of a probable SUDEP should of course be not one of foreseeing only and above all an ominous perspective to the child’s life, as well to that of the family. Thus, especially in cases when compliance is poor, and the seizure frequency is high (as expected), the relationship between responsible medical specialist and the family of the child, might become enormously complicated. A bad compliance and a precarious therapeutic alliance beyond all doubts need the question to be addressed sensitively; it might be necessary to hold closer contacts with the child family and to include the role of the psychologist in specific cases.

SUDEP is defined as a sudden, unexpected, witnessed or unwitnessed, nontraumatic and nondrowning death in a patient with epilepsy, with or without evidence of a seizure and excluding documented status epilepticus, in which postmortem examination does not reveal a toxicological or anatomical cause of death (1). Differences between the incidence of SUDEP in adult comparing to the incidence in children are obvious; in adult figures ranging from as low as 0.35 per 1000 patient years to as high as 9.3 per 1000 patient years are given from several sources (2, 3). On the other hand, figures regarding the incidence of SUDEP in childhood are far smaller than the over mentioned, ranging from a minimum of 1.1 per 10000 patient years to a maximum of 4.3 per 10000 patient years (4, 5, 6, 7).

Be it for a rarity, like the SUDEP in childhood as it is suggested from the figures above, it seems sometimes logical not to enter at all the discussion of a sudden death to an otherwise already traumatized family, because of the medical and economic impact of the epilepsy, as well as of the stigma that surrounds the suffering person from the disorder itself (8). Nevertheless, the issue has very strong moral, legal and medical implications, and the probability of a sudden death needs a discussion with the family of the child, although the timing and the communicative form remains still a controversy.

The frequency of the seizures is by large one of the most often discussed and implicated factors that lead to SUDEP, in adults as well as in children (9). Polytherapy has been also implicated as a risk factor for SUDEP, although the usage of two or more anti-epileptic drugs (AEDs) might as well indicate the poor seizure control (10, 11).
Long duration of epilepsy is another risk factor, and when discussing mortality in general (not strictly that related to SUDEP) authors consider that children with symptomatic epilepsy have a 22-fold increased mortality risk compared to the general paediatric population (12).

Regarding the death mechanisms involved during an epileptic seizure, the mostly discussed and accepted is the cardiac arrhythmia, in the form of an ictal bradycardia, ictal tachycardia or of an asystole (13). Postictal central or obstructive apnea has been reported (although in adult patients), and hypoxemia together with an increase of carbon dioxide partial pressure have been registered (14, 15). Apart from the multiple usage of antiepileptics (polytherapy) as a risk factor per se, carbamazepine solely has been blamed as a potential risk factor for SUDEP, especially in cases when high plasmatic levels of the drug were found (16, 17). Through suggesting the role of insufficient brain serotoninergic activity, some authors have even raised the hope that SSRI medications might in the future be an option for preventing SUDEP (18).

2. DISCUSSION

Debates related to the truth-telling and disclosing all details of a diagnosis, the prognosis related herein, risks and unforeseeable events that might overcome during the natural history of a chronic disease are as old as the Plato philosophical meditations in his Republic: “...Truth should have a high place among the virtues, for falsehood, as we were saying, is useless to the gods, and only useful to men as a medicine”, written around 360 B.C. (19). Anyway ‘falsehood’ as a ‘medicine’ might have some philosophical justifications, but from the medical point of view the growing trend is toward a more incisive truth-telling position, and this trend has had as well its historical slope (20).

Appleton has mentioned in details all important reasons why families need to know about the risk of SUDEP (12). According to him, premature death might occur in epileptic patients; SUDEP being a rarity is nevertheless not entirely predictable and compliance with antiepileptic medication is an issue; ‘sensationalist’ sources such as mass media will confuse even more families through offering non-professional or even erroneous information; and families have an inherent right to get all necessary information regarding the disease of their children, prognosis and risks related to the latter. albeit we mentioned above a clear trend toward truth-telling and disclosing a diagnosis, when questioning about the way professionals disclose the risk of SUDEP to their patients, some authors found that more than 60% of neurologists discussed the issue only with ‘very few of the patients’ (21). Pros and cons about disclosing the SUDEP possibility and the risk of the event have also been argued but till now no consensus position seems to have been validated (22).

The event of a child’s death is extremely traumatic for the Albanian culture. Logically such an event will provoke depressive reactions and even suicidal ideations; such reactions are described in Albanian epic songs like Ajkuna cries for Omer, dating several centuries before. In the latter the mother curses the moon, blaming the celestial object for the death of her son; Albanian versions of the epic song, actually forming part of the remote myth- eical epos of Balkans, are available online (23).

Acknowledging the exceptionality of SUDEP, clinicians however have to take into account the fact that first, it may happen and therefore the probability of the event, although remote, has to be addressed before the fatal seizure; second, the mere fact that a child is affected from epilepsy presents a huge psychological burden to the family, creating an abnormal parenting stress (24); and overreacting in these conditions seems to be a normal emotional response; third, professional advice is important since as already mentioned above, lay or non-professional information might be extremely dangerous.

Informing the families has to be made professionally (25). In our facility we offer to all patients and their relatives brochures with simple explanations and written in a lay terminology. Although terms like ‘death’ and ‘fatality’ are rather avoided in the brochures, the gravity of the diagnosis is of course mentioned, as well as the probability of long-term disability, together with the possible and future necessity for intensive care or intubation in cases of refractory epileptic status (such a situation is as well described in details); all these explanations leading to a better awareness of the gravity of seizures and of their possible complications. On the other hand, during the first monthly visits (patients registered anew to our facility perform monthly visits during the first semester), the specialized staff offers verbal explanations on how to cope with a seizure; by the third visit parents are generally aware that a generalized seizure can be fatal or cause irreversible damage.

Discussing the nature of an epileptic fit, sharing the family experience and projecting simple video explanations to them, are all very helpful in a setting where iatrogenic damages, such as tongue extraction or teeth breaking during awkward attempts to interrupt an apnea that normally accompanies a seizure (26), are extremely common, even among Albanian families living since years with epileptic patients. Recently an Albanian casuistic has been made available, with an unlucky case of infant death during a ‘crying spell’, when an overzealous and non-professional external chest massage caused myocardial contusion and cardiac arrest (27). In a small-scale descriptive study, a group of Albanian pediatricians and of other specialties has found difficulties in the families’ understanding of ‘epilepsy’ as a disorder: thus, 31% of the parents interviewed were feeling unsure; and another 7% as hopeless (28).

When coming to the preventive measures, we initially advised night supervision for the first trimester to all patients coming anew to our facility; but we strictly conceived such supervision merely as the presence of another person sleeping in the same room. Contrarily to what is suggested from other studies (29), to our experience, the option of listening devices in the child bedroom was found psychologically as aggravating the overall already tense familiar environment, and a possible risk factor for parental burn-out. Nevertheless, since sleep and SUDEP have been almost unequivocally interrelated,
there is a straightforward need to consider the risk of such a complication during the night hours (30). Albeit there are no definitive positions, we approach such a risk through (a) trying to raise the awareness of the relatives (families), even with the help of a person supervising and sharing the bedroom, (b) securing a better therapeutic coverage during the night hours, mainly through advising a higher evening drug dose; such a therapeutic position has been advised and found helpful from other authors as well (31).

3. CONCLUSION

To our opinion the disclosure of professional and timely information to the right interlocutor (parent and/or guardian) is a necessary act that will ensure a sound therapeutic alliance and a better pharmacological compliance, as well will ensure the moral and legal coverage to the treating clinician. We never discuss initially the concept of ‘death’ as an issue related to the diagnosis of child epilepsy; the newly registered patients are submitted to monthly controls for the first semester, and therefore, by the end of the third month, one of the relatives (parents) has already get the necessary information on the risk of a probable SUDEP situation. Our experience showed that a step-by-step approach and a gradual informing were helpful and psychologically acceptable from the parents or other relatives, taking care for the child suffering from epilepsy.

Notes: The authors declare that they have no conflict of interest. No external source has funded the present paper.

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