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Reprint of "Ethical issues with artificial nutrition of children with degenerative brain diseases"☆,☆☆,★

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ABSTRACT

This report highlights viewpoints of the authors and comments from the auditory at a workshop, held during the 14th international Congress on neuronal ceroid lipofuscinoses (NCL) in Córdoba, Argentina, on ethical aspects of artificial nutrition in children with degenerative brain diseases. The discussion centers on what constitutes the best interest of a patient whose personality was immature before the onset of the disease, who has become demented during its course and is unable to communicate his/her own positions and desires. There is wide consensus that in a child with advanced disease who cannot be fed naturally, decisions to withhold nutrition or to institute or stop artificial nutrition, should only be made by parents (or their representatives) who are adequately prepared on an intellectual and emotional level. We try to show that such decisions are highly individual but can be made in a rationally and emotionally acceptable way after a careful and prolonged dialogue between families and professionals. A checklist summarizes important considerations. This article is part of a Special Issue entitled: "Current Research on the Neuronal Ceroid Lipofuscinoses (Batten Disease)".

1. Introduction

The neuronal ceroid lipofuscinoses (NCL) are a group of genetic incurable degenerative brain diseases that constitute a relatively frequent prototype of a condition associated with dementia, complete helplessness and inability of communication at a young age [1]. While dementia, which denotes a loss of multiple higher cortical functions in a previously well-functioning person, is regarded as a public health priority by the World Health Organization [2], there is still little awareness of childhood dementia which occurs in a number or rare and mostly genetic brain diseases. Other disorders that develop problems similar to NCL during childhood or adolescence include the mucopolysaccharidoses and mucolipidoses, Niemann–Pick disease, certain leukodystrophies and basal ganglia degenerations. Such conditions raise a number of ethical issues, some of which are similar to those in the end-of-life care of elderly persons [3], while other topics are specifically related to the young age of patients (Table 1).

A frequent problem during the latter course of a degenerative brain disease in a child or adolescent is the inability of eating and drinking. With the availability of various methods of artificial nutrition, several questions arise regarding their application in children with degenerative brain diseases (Table 2). Compared with the growing knowledge and awareness of such diseases in recent years, little medical literature deals with these questions [4,5], although some principles regarding children with severe brain damage have been expressed many years ago [6]. It is clear that other medical therapies aiming at the prolongation of life in the end stage of an incurable brain disease, such as respiratory support, are subject to analogous deliberations.

Professionals caring for children with degenerative brain disease are in need of principles that are applicable in practice when the question of

☆ This article is part of a Special Issue entitled: "Current Research on the Neuronal Ceroid Lipofuscinoses (Batten Disease)".

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Table 1
Reasons that make some ethical issues specific to demented children.

- The patient's personal autonomy is not yet fully developed.
- The patient has had no chance yet to conceive and fix a consistent conviction regarding questions of vital importance.
- As dementia in childhood is rare, relevant knowledge and experience are scarce.

tube feeding arises. Clear concepts should create a feeling of security in caregivers and avoid traumatic irritations through the interfering of persons not directly involved.

2. Origin of presented material

This article is a report of issues discussed at a workshop where the authors presented their views on aspects of life support through artificial nutrition for children in the end stage of a progressive brain disease. Members of NCL patient organizations participated in the discussion. For many critical questions a consensus was reached, mostly on the basis of intensive personal experience of families and medical professionals.

3. Issues dealt with at the workshop

Discussions centered on a list of questions where a need was felt for analysis of frequently encountered medical situations and for solutions or guidelines to management (Table 2).

In the following, we treat these questions and offer some practical guidelines for handling such issues in young persons with a degenerative brain disease.

3.1 What are medical indications for an artificial nutrition?

Artificial nutrition and artificial hydration are medical treatments associated with significant problems and should never be instituted without an unambiguous medical indication. Although the decision to start such treatment is ideally based on a dialogue between the patient's family and their physician (see below), it is the latter's duty to formulate the indication and put it down in writing with simple clear words. Such a drastic medical measure as artificial nutrition should not be started as "the normal thing to do" in progressive dementia.

The indication may be easy to determine in patients with swallowing problems but who are otherwise in relatively good condition. Insufficient nutrition in such patients may lead to avoidable additional morbidity and unnecessary suffering. The indication is much less clear in far advanced stages of cerebral decline, as improvement of the quality of life through artificial nutrition may not be convincing at this stage. Quality of life, however, is a very variable notion that only can be assessed regarding the individual situation. As a rule this is only possible for parents and for physicians thoroughly acquainted with the medical history.

Table 2
Questions related to artificial nutrition in children with degenerative brain disease.

When a child in the later stage of disease has become unable to be fed naturally:
- What are medical indications for an artificial nutrition?
- When is prolonging life by artificial nutrition in the best interest of the patient?
- Can artificial nutrition be withheld or terminated in certain situations?
- If artificial nutrition is indicated, what methods should be used?
- By whom and how should decisions be made?
- What are the psychological, social and legal implications of such decisions?

3.2 When is prolonging life by artificial nutrition in the best interest of the patient?

There is little doubt that decisions of vital importance should be made in the best interest of the patient, but difficulties may arise as to what constitutes the patient's best interest, particularly in a mentally incapable, non-communicative child with severe brain damage [7]. Sometimes, arguments outside the realm of empathy and the presumed well-being of the child are put forward, such as "sanctity of life" or other concepts colored by religious, cultural, and racial or ethnic perspectives. Such arguments are rarely emphasized by persons who have been intensively confronted with the situation of such children. There seems to be a wide consensus that ethical questions of the nature discussed here should be settled on a highly individual basis and not according to rules or directions of outsiders.

It must be emphasized here that feeding against the patient's wishes is widely regarded as an assault on personal integrity. In this respect, the presumed wish of a demented patient (or the wish reasonably projected on the patient) not to be fed artificially is particularly weighty and has been subject of intense discussion in demented adults [8].

3.3. Can artificial nutrition be withheld or terminated in certain situations?

When, for whatever motive, artificial nutrition has been instituted, a situation may evolve with good reasons for termination. After prolonged observation of the patient, parents and physicians may come to recognize that continuation of artificial nutrition will be without benefit to the patient and will prolong life unnaturally. In this situation, parents, who are the only ones responsible for their child's welfare, may be under psychological pressure towards continuation of artificial nutrition.

If a decision to withdraw or withhold treatment that prolongs life is considered to be in the best interest of the patient, it does not contradict prevailing laws in most countries. This implicates that it can be in the patient's best interest to accept death as a consequence of the decision. These deliberations should be touched upon and be adequately documented already at the very first consultation where questions of nutrition come up.

It should be noted that the concept of artificial nutrition comprises the supply of both, food as well as water. Medical experts expressed concern that withholding food but continuing the supply of water would lead to a particularly poor physical condition and undesirable prolongation of life. In a patient dying from a cerebral condition as discussed here, keeping the oral mucous membranes moist is thought to be adequate palliative care.

3.4 If artificial nutrition is indicated, what methods should be used?

Clinical judgment must be used to determine if nutritional support will improve the patient's well-being. Evaluating growth and the gaining of weight of a severely handicapped child on the basis of the usual pediatric charts is not useful. In immobile adolescents, obesity provoked by relative over-feeding sometimes creates unnecessary difficulties of handling the patient.

Once a clear decision has been made for tube feeding, the help of a gastroenterologist with pediatric experience must be sought. Percutaneous endoscopic gastrostomy is a good solution and feasible in many parts of the world [9]. If this service is not available, naso-gastric tubes are an alternative that frequently works for prolonged periods of time without serious complications.

The type of food used for artificial nutrition via tubes deserves critical examination. As these neurological patients usually do not have an intestinal disease, food composition should be close to a regular diet. Food can be prepared at home using mechanical homogenization. This is not only economical but also avoids much discomfort that is associated with industrial preparations. Erroneously, unphysiological high-