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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)".

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### 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 of rare and mostly genetic brain diseases. Other disorders that develop problems similar to NCL during childhood or adolescence include the mucopolysaccharidoses and mucopolipidoses, 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

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**Table 1**

Reasons that make some ethical issues specific to demented children.

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- 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.

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- 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-

calorie preparations are frequently given to patients with muscular atrophy due not to malnutrition but to motor inactivity. Caloric requirements should not be prescribed on the basis of healthy children of the same age. Other details, such as the schedule of feeding in individual patients, are beyond the scope of this discussion.

### 3.5 By whom and how should decisions be made?

The nature of the disease and the present condition of the patient must be totally clear for those who take responsibility for decision-making. Is it the hopeless end stage of an incurable disease where nothing can be gained but a prolongation of life? Or is it a condition where improving the nutritional status will, with some probability, lead to less discomfort and more quality of life? This may be the case, for example, in a patient with serious difficulties of swallowing due to neurological dysfunction. Such patients may be alert but may try to avoid the life-threatening experience of choking because of aspiration of nutrients, which they express by clenching their teeth.

As the patient cannot decide for himself/herself because of legal immaturity and the deficiencies caused by disease, others have to carry the difficult burden. While generally, according to the 1989 international Convention on the Rights of the Child, rights and responsibilities are shared between children, parents, researchers, and the state [10], in the context of the present discussion, it is usually *only the parents* who are qualified to decide what is in the best interest of their child.

Qualified decisions on whether to start or stop methods of vital support in their own child, however, require an enormous amount of factual knowledge, intellectual judgment regarding the past evolution of the disease, the present medical condition and imagination about the future, psychological and emotional strength, as well as some sense of how one's own feelings and those of others react with regard to the decision at stake. This situation obviously requires help, as becoming competent for the decision-making is a prolonged process.

Supporting parent autonomy and competence in decision-making regarding treatment is complex. Making parents understand the medical aspects is physicians' duty, but the necessary teaching and learning can happen in many different ways [11]. Attitudes of medical professionals in such a process can be "paternalistic", which still seems to be accepted in some areas, and will tend to ease the burden on parents by strongly suggesting what is to be done according to authorities. The other extreme attitude is an "objective approach", where the physician presents the different choices, their pros and cons, without giving recommendations, leaving the decision-making exclusively to the parents. This type of "objectivity" neglects an inherent cruelty of forcing decisions from inadequately prepared parents. How differently families react to the necessity of vital decisions for their children has been studied in the high-stress environment of the pediatric intensive care unit [12], while little has been published regarding slowly deteriorating children at home.

Decisions should be the result of a carefully led process of deliberations and dialogues between the family and their physician(s). As questions related to stopping food supply are emotionally disturbing, working decisions are only achieved in an atmosphere of trust and empathy that requires a relatively long period of time. Parents must be informed about the condition of their child, options at the different stages of the disease discussed, and unrealistic positions and futile interventions discouraged. Psychosocial support may have to be organized.

As a consequence of the complexity of problems, an interdisciplinary team should be established, which is led by the professional most familiar with the patient's needs and in which the family is a significant member.

### 3.6. Documentation

An adequate possibility to document decisions on life support is making an entry in a form that is frequently used in adults who

want to avoid futile attempts to keep them alive in a hopeless situation, a so-called DNR (Do Not Resuscitate) form.

### 3.7 Psychological, social, legal and financial implications of such decisions

Artificial feeding of a young child with vanishing brain function, who would die without this measure, will make the patient look less disturbing than when left to progressive malnutrition. With good care, the patient may continue to live in this way for many years and may reach the age of adolescence and adulthood. The parents will grow older themselves, becoming increasingly unable to care for their child, who eventually will be transferred from the family to a nursing home. Such deliberations may help families to develop a more realistic view of their beloved one when they feel an inadequate propensity to opt for life-supporting measures.

A more "natural" attitude towards the fateful experience with a hopelessly sick child may help to reach what some have thought to be one of the aims of management: dying with dignity.

The role of patients' associations can be equivocal in the context of pending decisions about artificial nutrition. On the one hand, advice can be gathered there from families who have experience with the subject. On the other hand, individual attitudes towards highly emotionally loaded questions may vary widely and may contribute to the confusion of parents.

"Quality of life" in children with severe diseases is seen very differently by families. While some may think that a child who is not communicative and not running around has a low quality of life, others will value a small remaining ability to show emotional reactions as evidence of a significant quality of life. While professionals should try to understand such feelings, they should not interfere with them and respect them as an expression of legitimate human individuality.

### 3.8 Legal aspects

In a standard situation, the complex and vital questions discussed here only concern the patient, the family and (personally involved) physician(s). A simple documentation of important decisions and the arguments supporting them is advisable. Involvement of outsiders not acquainted with the individual case (such as ethical committees, lawyers, judges) may not be necessary if it is in line with national legal regulations. The inclusion of outsiders in the process is advisable in situations with dissension between parents, between parents and physician, or when there is uncertainty about the custody of the patient. In such cases, a person working in the institution where the child is primarily being cared for may be a preferred consultant, sometimes an institution's ethical committee.

### 3.9. Financial aspects

The costs of artificial nutrition are highly variable depending on the type of food selected for tube feeding. Home-prepared homogenized meals are not only advisable in most cases for medical reasons mentioned above, but they are also much cheaper than artificial formulas. Other components of the financial burden of artificial nutrition are the degree of involvement of ambulatory palliative care teams or other personnel. The financial strain on the family will depend on the local social security systems. It should be noted that, deplorably, under conditions of lack of time, tube feeding is sometimes preferred to time-consuming attempts at natural feeding.

## 4. Recommendations

In any situation where initiating and terminating artificial feeding of a child with degenerative brain disease is contemplated, several points must be clarified, as shown in the check list below (Table 3).

**Table 3**

Check list “Artificial feeding of children with degenerative brain disease”.

- 
1. Is there an unambiguous medical indication to start artificial nutrition?
  2. Does the family understand the medical situation and the potential consequences of artificial nutrition (prolonging life unnaturally without benefit to the patient)?
  3. Does the family know that artificial nutrition can be stopped, when the indication for it is no longer valid, and what the consequences of a stop will be?
  4. Have these points been adequately documented?
  5. As artificial nutrition is an active medical treatment with significant consequences and risks, who is going to take the lead for the long-term management?
  6. Does the family need additional psychosocial support?
  7. Is the financial basis of management clarified (insurance and other sources)?
- 

## 5. Conclusion

In a child in the advanced stages of a degenerative brain disease who cannot be fed naturally, decisions to institute artificial nutrition, to withhold nutrition, or to stop artificial nutrition that was started at an earlier stage, can be made in a way that is rationally and emotionally acceptable. Such decisions require taking into account a number of considerations that have been discussed at the workshop and highlighted in this article.

## Transparency document

The [Transparency document](#) associated with this article can be found, in the online version.

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