

UPDATE

Treatment of lumbar and intrathoracic meningocele: bioethical implications

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Abstract. Myelomeningocele is a congenital malformation caused by a developmental defect of the spinal cord structures. The exact cause is unknown, but different factors have been involved including radiation, malnutrition, drugs. Myelomeningocele can develop at any point in the spine, but the lumbosacral region is affected in over 75% of cases. Chest X-rays and computed tomography study are mandatory to reveal tracheal malformations or associated anomalies of the ribs. Treatment of myelomeningocele must be multidisciplinary and involve at the same time neurologists, radiologists, neurosurgeons, thoracic surgeons, bioethical experts and take care of the child and also of the family. Some experiences concern the possibility of an in-utero correction of myelomeningocele, in order to avoid serious and progressive damages to the nervous system. Given the improvement of myelomeningocele management, the quality of life is nowadays more acceptable than in the past; however, some severe forms of myelomeningocele cannot still be corrected: in these cases, a “non-interventional” approach may require a form of passive euthanasia that should be discussed and approved with and by parents and any dissent of the parents must be respected and considered reasonable. The choice of a “non-intervention”, which should be guaranteed to all the people capable of self-determination, is not however so immediate and direct in the case of the minor: the dissent expressed on his behalf by the parents or legal representative may be ethically difficult to be accepted. In this case, the best interest of the child must prevail as the goal of any therapeutic choice. (www.actabiomedica.it)

Key words: Meningocele, Lumbar, Intrathoracic, Bioethics, In-Utero Surgery

Introduction

Myelomeningocele is a congenital malformation caused by a developmental defect of the spinal cord structures (i.e. meninges and vertebrae) during the embryonic period, mainly between the third and fourth weeks of gestation, and is the result of a failure in the closure of the posterior neural tube. Its incidence in recent years is decreasing due to the possibility of carrying out prenatal diagnosis and the possible

interruption of pregnancies of affected fetuses, as well as the increase of folic acid assumption in the first weeks of pregnancies (1). The exact cause is unknown, but it is likely that many factors may be involved, including radiation, malnutrition, drugs consumption. These environmental and nutritional factors, however, act on a genetic predisposition (still unknown), demonstrated by the fact that the risk of recurrence is 3-4% after a first child with myelomeningocele and rises to 10% after two affected children. It is now widely

accepted that feeding with folic acid, which starts two to three months before conception and which should be prolonged up to the twelfth week of pregnancy, reduces the risk of myelomeningoceles in conceived by 50-70%. To this aim, in the USA, foods for pregnant women such as bread, pasta and cereals have been enriched with folic acid (1,2).

Diagnosis

The prenatal diagnosis of myelomeningoceles is usually made directly by ultrasound and indirectly by evaluating alpha fetus protein levels in amniotic fluid and/or maternal serum. High levels of alpha fetus protein at 16-18th week of gestation have a high predictive value of neural tube defect and subsequent disorders of the central nervous system (3-6). At the ultrasound scans, myelomeningocele is found mainly in the lumbar region and usually occurs as a cyst covered or not with healthy skin. The vertebral arches are absent and the posterior peduncles of the vertebrae are displaced laterally causing an increase in size of the spinal canal. Although the anterior part of the spinal cord is well structured, the dorsal cord is completely disorganized with nerve tissue mixed to fibrovascular elements.

Symptoms caused by myelomeningocele are different, and depend on the level of the lesion. For example, if it is located under the sacrum, the child may be able to walk, but if it is higher, beyond the second lumbar vertebra, it is almost certain that he will have to spend his life on a wheelchair for the involvement of nervous roots of the nerves of the legs(7). Myelomeningocele can actually develop at any point in the spine, but the lumbosacral region is affected in over 75% of cases. When the lower sacral region is affected, the child may then walk, but will suffer from urinary and faecal incontinence with anesthesia in the perianal area. Upper lesion levels involve flaccid paralysis of the lower limbs, reduced or absent sensitivity to touch and pain and a high incidence of postural defects with dislocated hips and distorted feet. Up to 80% of children with myelomeningocele also have hydrocephalus (Arnold-Chiari malformation). Other important clinical consequences of myelomeningoceles concern the intestinal and urinary tract. Children

with myelomeningocele present in most cases malformations such as bladder-ureteral reflux and hydro-nephrosis, which cause frequent urinary infections, nephropathy and, in the most severe cases, chronic renal failure (8).

Malformations of other organs can be present. In a recent paper reporting the review of chest X-rays of 87 patients with myelomeningocele, it was reported that 31 (36%) had short tracheas. This degree of association of short trachea with myelomeningocele, the frequency of myelomeningocele, and the number of surgical procedures performed suggest that special attention to the short trachea is warranted in myelomeningocele patients because of the risk of accidental bronchial intubation and subsequent sequelae. Malformation of the twelfth rib was present in 23 patient: 12 (14%) with complete aplasia and 11 (13%) presented a hypoplasia (9-11).

Moreover, computed tomography study may show cystic mass extending through enlarged intervertebral foramen. Cystic nature of this mass, in this clinical setting, is virtually diagnostic of lateral thoracic meningocele (12).

Symptoms

The child with myelomeningocele presents a series of problems ranging from flaccid paralysis to hydrocephalus, urinary and faecal incontinence, chronic kidney disease and renal failure. Thoracic meningoceles often are asymptomatic or produce radicular intercostal pain. Differential diagnosis should include mediastinal or spine tumours; in the management of patients identification of early parathormone (PTH) changes may have a predictive value on thyroid and parathyroid involvement, as well as videomediastinoscopy, which is mandatory for a correct diagnosis (13,14).

Treatment

The treatment of myelomeningocele must be multidisciplinary (involving neurologists, pediatricians, radiologists, neurosurgeons, thoracic surgeons, bioethics experts) and take care of the small and also of

the family. The care coordination should be entrusted to a pediatrician. For the treatment of myelomeningocele protocols are based on the type and severity of the lesion, on the basis of which the neurosurgical interventions are modulated.

The most frequent decision is to proceed with myelomeningocele correction as soon as possible. Nevertheless in thoracic myelomeningocele, a minimally invasive procedure such as uniportal VATS (15-17) and transcervical approach could be required in symptomatic cases, usually with total remission of the pain.

Some experiences concern the possibility of intervening on the repair of myelomeningocele in utero, in the hope of avoiding more serious damage to the nervous system. (18,19). In fetuses affected by myelomeningocele, movements in utero are present in the early weeks of pregnancy, then gradually decreasing until it disappears after the birth at term, suggesting a progressive loss of neuromotor function.

In-utero interventions, however, raise serious technical and ethical problems, because in prenatal surgery there are two patients, the mother and the fetus, and the surgery risks involve a person, the mother, who is not directly physically involved. Furthermore, experience is currently limited to be able to satisfactorily define the advantages and possible complications of this procedure. (20).

When the surgery is performed after birth, specialists take care of the child and start the multidisciplinary treatment program. The neurosurgeon and the neurologist will evaluate the level and severity of the lesion with specific investigations and the need for ventricular derivation intervention and timing. Bladder dysfunction and urological pathology require a series of radiological and functional investigations in order to prevent severe urinary infections and irreversible renal damage. Specific interventions implemented to limit the consequences of the neurological injury of the lower limbs can allow the recovery of muscle activity and to correct the malformations that may be present since birth (e.g. subluxation or dislocation of the hips and the distorted feet). These interventions improve the locomotion abilities, achieving a greater patient autonomy, as well as the cognitive functions.

The intelligence of children with myelomeningocele develops according to the severity of brain impairment,

which is mainly related to hydrocephalus. In 40% of subjects with normal intelligence, however, deficits of psycho-intellectual abilities and attention skills, memorization in school learning are demonstrable. Advances in medicine have led to the survival of newborns with myelomeningocele; mortality, which was 100% in the 60s, currently ranges from 10 to 15% in the earliest ages of life, with higher mortality rates before 4 years of age. Those who survive have a very long life expectancy if complications do not occur (brain infections, valve mal-function, respiratory failure due to deformity of the rib cage, etc.). Rarely, sudden death may be caused by compression of the medulla oblongata.

The quality of life of the subjects affected by this serious and complex pathology is better than in the past for the implementation of health supports, even if myelomeningocele is related to several chronic consequences (21-25). The consideration of the difficult path that awaits the subject affected by myelomeningocele, sometimes without the prospect of achieving a full autonomy, and also of the enormous costs for the society of assistance to these patients, has led the UK to develop strict criteria for the exclusion from treatment of those affected by the more severe forms of myelomeningocele. These criteria include paralysis of the lower limbs, thoraco-lumbar and thoracolumbar sacral lesions, kyphosis and scoliosis, cardiac, encephalic and gastrointestinal defects, and finally giant hydrocephalus (21,26). However, in many centers, physicians continue to aggressively treat all those affected by myelomeningocele and many of them manage to achieve high levels of economic autonomy with full integration into the working world and society.

Myelomeningocele causes very serious disability and requires continuous treatment, which however are effective because they allow in most cases not only survival but also a discrete or even good psycho-physical recovery. The treatment of myelomeningocele and its complications therefore does not represent a condition of therapeutic obstinacy (as defined by the Deontological Code) and the physician is obliged to evaluate the most useful treatment for the patient, especially when his survival will not cause discomfort and disturbance to the family.

In case of opposition from the legal representatives to the necessary care of minors and the incapacitated, the physician must request the intervention of

the competent Judge, as the decision to not operate the child with myelomeningocele would be resolved in the voluntary suppression of a newborn, while the decision to operate the child cannot prefigure a case of therapeutic obstinacy. Surgery, in this case, is an ordinary and not extraordinary therapeutic measure and the affected subject, as a result of those cures, can not only survive, but develop physically and psychically. The child with myelomeningocele has “a life project” which will be very difficult but inalienable.

Bioethical Considerations

The management of myelomeningocele raises the question about the opportunity to undertake intensive care in the case of severely pathological infants. It is necessary to evaluate which type of treatment should be reserved for these children for whom, given their condition of serious illness, there is currently no hope of recovery, since no cure allowing them to return to normal health conditions is available, neither from the physical point of view nor from the mental point of view. Moreover, these cases are increasing, because of the improvement in medicine and technology that have greatly improved the prognosis of many congenital diseases. The availability of increasingly advanced and sophisticated care techniques opens the question of which therapies should be undertaken.

In general, the questions raised can be formulated as follows:

- a) whether or not to initiate resuscitation on a newborn with a poor chance of survival and with a high probability of permanent severe deficits;
- b) whether or not to perform surgery to correct abnormalities in subjects destined to a short and highly compromised life;
- c) whether to continue or discontinue intensive care once it is undertaken, but only to provide basic care to young patients (27-30).

End-of-life decisions, which are very demanding when they are taken for a patient in conditions of particular fragility, as in the case of myelomeningocele,

must be taken in his best interest. The main problem concerns the right to refuse treatments that unnecessarily prolong the life of the newborn when, in the opinion of the doctors, these are useless or useful only to prolong the process of dying in a definitive way.

The different evaluation of the decision to treat or not treat the newborn depends on the value attributed to life: in the perspective of a “pure” defense of life (considering it as a value regardless of its quality), resuscitation maneuvers are not only lawful but also strongly recommended. In the opposite perspective that instead focuses attention on the quality of life, treating the subject with myelomeningocele means not giving her the chance of life of “quality”, but rather guaranteeing only a further survival time. Given the serious disease condition of myelomeningocele, surgery with uncertain outcomes may not be the best possible solution. In relation to the decision to be taken in such difficult cases, it can be said that there is a broad consensus on the opportunity to avoid any therapeutic obstinacy and on the duty to spare the child unnecessary suffering, even if the use of further drugs could accelerate the death.

While it is easy and rewarding to assist patients who may benefit from medical-technological approach, it becomes much more difficult and problematic to make decisions about a child who is dying or who is or will be severely handicapped, with serious injuries incompatible with an acceptable life or continuous suffering (31). In the case of the newborn affected by myelomeningocele there would not be a real indication for the proposed surgical interventions since it is not expected to derive benefit for the subject in its entirety. The decision to suspend treatment appears then as the best possible alternative.

It should be noted that limiting to basic care, or containing therapies is not equivalent to practicing euthanasia: suspending invasive and disproportionate treatments compared to the realistically achievable result, while ensuring basic care, does not mean procuring death, but means accepting death as a result of the particular disease condition.

In Italy, the criminal discipline related to euthanasia can be traced back to three criminal provisions of the Penal Code and a reference of a constitutional nature. The Penal code Defines in article 575, the

crime of voluntary homicide, if the death event is carried out through omissive conduct; with the article 579, on the other hand, the murder of the consenting party is incriminated, albeit with reduced penalty compared to the provisions for voluntary homicide in general; article 580 defines the instigation to suicide as a misconduct. In this way, Italian system is currently held to be the condemn all the forms of active euthanasia, even if carried out with the consent of the person concerned; also non-consensual passive euthanasia may be regarded as a transgression.

In this sense, this may contrast the law reference of the Constitution, especially article 32 (paragraph 2), which establishes that no one can be obliged to a specific health treatment except by law, and, and article 13, which protects the freedom of the individual (including the right to the refusal of treatment by the patient). Therefore, consensual passive euthanasia would certainly be licit or, even, it would not even be considered a form of euthanasia; in fact, it would represent only a manifestation of the respect - from the doctor - to the right of not be cared by the severely sick individual.

Different problems, therefore, given the absence of a specific discipline - and in any case of normative data from which to draw unequivocal indications on the point - arise, in particular, with reference to cases of non-consensual passive euthanasia, or on subjects incapable of expressing their own will (32,33). A particular hypothesis of this wider problem is the situation of newborns suffering from severe malformations, such as myelomeningocele. In the case in which the patient is a minor, the subjects in the first instance who are holders of the guarantee obligation (protection) are not the doctors, but the parents. These, according to the articles 30 of the Constitution and 147 of the Civil Code, have the duty to maintain, instruct and educate their children and also to preserve their health. It is therefore necessary to establish whether, in the event of the child's death, the dissent to the intervention, expressed in the name and on behalf of the child by the parents, can lead to their responsibility for murder by omission (Article 575 of the Italian Penal Code), or if the decision to not intervene, expressed by the legal representative, can be equated with a disagreement expressed by the same subject and, therefore, based on

art. 32 of the Constitution, is an expression of the constitutionally guaranteed right of not being treated and not a crime.

In cases of serious illness, a conflict may arise between the parents or the legal representatives, and the doctors. When the parents, informed about the risks and benefits of a specific intervention or therapy, decide to continue the treatment, physicians cannot oppose, especially when the possibility of saving or improving the child's health conditions. In this hypothesis the doctor has the duty to take action, except that the treatment must be considered harmful to the child, or without any benefits (29). If the parent does not agree with the decision of the physician, in this case they have to contact the competent authority, who will assess the parents' decision regarding the minor and take the appropriate measures, in the interests of the minor. This is guaranteed by Penal Code articles 330, 333, 336 c.c. and sometimes the art. 403 c.c. which provides for the possibility of public authority intervention in favor of minors. In particular, the judge can rule, in extreme cases, the cessation of parental authority, and issue a provision for the enforcement of the therapy.

However, it is not easy to define the boundary beyond which the physician's activity is transformed into therapeutic obstinacy and the difference between lawful medical treatment and therapeutic obstinacy is generally identified through the reference to the distinction between ordinary and extraordinary means of treatment; in this sense, "ordinary" not simply means the usual procedures, but the appropriate and proportionate interventions. The concept of therapeutic fury implies the loss of the benefit for the patient, the ultimate goal of medical activity. Parents, legal representatives of the child, are the subjects holding a position of guarantee and protection of their child: they also have the power to express consent or dissent to care, in the name and on behalf of the minor.

Surely parents must not be left alone, for the good of the child: the difficulty of a decision in cases of malformations of severe newborns, and a very poor prognosis, as in the case of myelomeningocele, is also accentuated by the impossibility of having the will expressed by the minor. The law must demand, in the presence of "weak" subjects, that the person designated

for their protection fulfills the task assigned to them, in a rational and reasonable manner (31). Therefore, the solution of such cases cannot be entrusted only to the parents. A dialogue with the physicians is necessary and, if necessary, the decision of a third party, expressed following a careful and rigorous evaluation of the costs and benefits, which takes into account primarily the outcomes of an intervention on the child and his life chances of social integration.

Conclusions

In the treatment of myelomeningocele, the progresses of medicine have been enormous: in addition to a drastic reduction in mortality, today we are witnessing the possibility of achieving a better quality of life than in the past. The choice of a not-intervention would, of course, involve, from a technical point of view, a conduct of passive euthanasia which should be discussed and approved when it is considered as one of the options and any dissent of the parents must be respected and considered reasonable. The right not to care, which is guaranteed to the person capable of self-determination, is not however so immediate and direct in the case of the minor and the dissent expressed on his behalf by the legal representative. Although in the adult the right to self-determination of the subject prevails, in the minor the best interest of the child must prevail as the goal of a therapeutic choice.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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