Presidenza del Consiglio dei Ministri

CONJOINED TWINS AND SURGICAL SEPARATION: BIOETHICAL ASPECTS

19th of July 2013

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Presentation

In this opinion the Committee dealt with a delicate, even though not frequent issue, which calls for deep bioethical reflection that might be of guidance in complex decisions at clinical level.

Even though recognising the complexity and variability of the single cases, the Committee tackles two main situations, with reference to new born babies/minors.

A first condition is the one in which the life of the twins is not in immediate danger, while the surgical separation, although technically possible, is highly risky for the life of one or both twins. The document highlights two principle lines of thought: the line of those who, referring to the value of human life, consider that in the measure in which an operation is not necessary and is disproportionate, it is not ethically justified; the line of those who on the basis of various arguments consider surgical separation ethically licit even when the risk is high, as long as it creates even slender hopes of success.

A second condition is that in which, on the basis of an objective clinical evaluation supported by empirical data, the certainty is manifested of the imminent and serious threat to the lives of both the twins. Different lines of thought are to be found in this context too: on the basis of different arguments some consider that the parents' choice of not intervening is justifiable, also in contrast with the doctor's clinical opinion; the wide majority of the Committee considers that before an appreciable and reasonable forecast of a slender chance of survival for one of the twins, surgical separation is dutiful, invoking the protection of life.

In the conclusions the document outlines a number of guidelines as a reference context to foster ethically complex decisions at clinical level, which refer to the promotion of research and the professional training of the healthcare operators, the importance of adequate information and psychological support for the parents, the duty to not intervene should experimental persistence be presented, the role of the ethical committee and the duty to maintain confidentiality.

The Committee maintains that in the case of adults, the wish of the twins must be considered identical to that of any other competent adult, even with reference to the choice of undergoing experimental treatment or of refusing therapy.

The opinion was coordinated and drafted by Profs. Salvatore Amato, Lorenzo d'Avack, Laura Palazzani, with substantial contributions from Profs. Bruno Dallapiccola, Adriano Bompiani, Stefano Canestrari, Riccardo Di Segni. Profs. Antonio Da Re, Francesco D'Agostino, Marianna Gensabella, Assuntina Morresi, Demetrio Neri, Monica Toraldo di Francia took part in the work group and contributed to the debate.

A precious contribution was given by the first-hand experience of Profs.: Bruno Dallapiccola (Scientific Director of the Paediatric Hospital Bambino Gesù of Rome and member of the National Bioethics Committee), Pierpaolo Mastroiacovo (President of the Ethics Committee of the Paediatric Hospital Bambino Gesù, Director of the "Central Office of the International Clearinghouse for Birth Defects Surveillance and Research" and "ICBD – Alessandra Lisi International Centre on Birth Defects and Prematurity", WHO Collaborating Centre), Pietro Bagolan (Director of the Department of Surgical

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The opinion was approved unanimously during the plenary session by Profs.: Luisella Battaglia, Stefano Canestrari, Bruno Dallapiccola, Antonio Da Re, Lorenzo d'Avack, Maria Luisa Di Pietro, Riccardo Di Segni, Romano Forleo, Silvio Garattini, Marianna Gensabella, Laura Guidoni, Assuntina Morresi, Andrea Nicolussi, Laura Palazzani, Monica Toraldo di Francia, Giancarlo Umani Ronchi, Grazia Zuffa. Profs.: Cinzia Caporale, Francesco D'Agostino, Carlo Flamigni, Demetrio Neri, Vittorio Possenti, Rodolfo Proietti, Lucetta Scaraffia expressed their approval at a later date.

The President
Prof. Francesco Paolo Casavola

I. Brief scientific description of the phenomenon

Definition

Conjoined[1] twins are united by one part or several parts of the body and may share apparatus and organs[2].

History

References to them exist in different cultures, from far-off ages. Numerous cases have been recorded (the first in England in around the XII century, in Venice in the XVI century, in Hungary and Bohemia in the XVIII century and in Siam in the XIX century) and scientifically analysed (the first was described in medical literature in Mexico in 1868). Many of them were considered as a 'curiosity' or a 'monstrosity'[3] and were all too frequently used for shows in the past.

Etiology

The etiology is uncertain[4]. There are two scientific theories explaining their origin with reference to an abnormal embryonic development: the *fusion theory* and the *fission theory*.

The fusion theory explains the conjunction of the twin birth making reference to the process of fusion, by means of which the two separate embryos partially fuse together. This theory does not explain the prevalent symmetrical conjunction of the twins (that is, always in the same part of the body or the so-called *mirror-image*), with the only exception of the phenomenon of the parasitic twin[5]: the chance fusion could result in an asymmetrical conjunction.

The fission theory, which is the most substantiated today, explains the phenomenon as a late and incomplete scission of the zygote; after fertilisation, the embryo does not complete its division. Initially only one, the embryo undergoes a late separation around the 14th to 15th day following fertilisation and is not able to complete the separation of the two inner cell masses. Consequently, instead of producing two embryos that are distinct and independent in their parts, sharing a single placenta, it produces two partially conjoined embryos. A small percentage of identical twins presents a single *corion* (placenta) and a single *amnios*. This means that the division of the

[1] The terminology 'Siamese twins' derives from the Chang and Eng Bunker twins (1811-1874) from Siam (today' Thailand). This is nevertheless a non-scientific term, even if widely used.

[2] O.M. Mutchinick et al., *Conjoined Twins: A Worldwide Collaborative Epidemiological Study of the International Clearinghouse for Birth Defects Surveillance and Research*, in "American Journal of Medical Genetics", Part C (Seminars in Medical Genetics), 2011, 157, pp. 274-287. The article gives the most extensive study of the phenomenon in the context of worldwide multi-centric research.

[3] G.M. Gould, W.L. Pyle (eds.), *Anomalies and Curiosities of Medicine*, ch. V Major Terata, W.B. Saunders, Philadelphia 1986, pp. 162-213.

[4] In ancient times and during the Middle Ages the birth of conjoined twins was said to be caused by: interventions of the devil, type of food, position of the woman during pregnancy.

[5] It is the phenomenon by which only a partially formed baby can join itself at the same part, for example head/head, or at a different part of the other twin's body.

embryo took place after the 9th day. These embryos risk giving rise to conjoined twins[6].

In the case of *conjoined twins* it is considered that the error in development occurs around the 14th to 15th day after fertilisation. This results in *monochorionic* and *monoamniotic* twins. The non completion of the separation of the precocious inner cell mass (*germ*) into two distinct embryos brings about the sharing of anatomical parts, organs, blood vessels etc., in a combination that can be extremely variable from case to case. These anatomical and structural differences not only classify the typology of the conjoined twins, but also affect their survival and the possibilities of successfully separating them surgically.

In order to explain the formation of conjoined twins hypothetically, one can refer to a partial 'splitting' of the longitudinal primitive streak (cranial-caudal). If this splitting takes place towards what is destined to become the cephalic end of the embryo, it should lead to the duplication of some parts that originate from this end (for example two heads, two chests etc.); if instead, the splitting takes place in the opposite caudal direction, duplications of the parts collocated along this direction arise (for example, double intestine, double liver, double pelvis, etc.). If the phenomenon of the right or left lateralisation with respect to the primitive streak of the collation of some organs is added to this process, the general picture of the clinical presentation of the conjoined twins will be complicated by the coexistence of further anomalies.

Typologies

The various clinical presentations of conjoined twins have given rise to different classifications. The most widespread differentiates twins on the basis of the conjoined parts of the body and the shared organs (with the use of the Greek suffix 'pagus' that means 'fixed'): *thoracopagus* twins, postioned one opposite the other and conjoined from the upper part of the thorax to the upper part of the abdomen; *cephalopagus* twins, joined from the upper part of the head to the navel, with two incomplete faces, two necks and the remaining parts of the body separate; *parapagus* twins, joined laterally, that generally share the pelvis, with heads and upper limbs separate and generally three legs: *omphalopagos* twins, conjoined at the level of the navel region and the low thorax, with a separate or conjoined heart; *craniopagus* twins, conjoined at the

[6] This is the thesis of Scott F. Gilbert, *Biologia dello sviluppo*, VIII, Bologna 2005, p. 333; M. Barbieri, P. Carinci, *Embriologia*, Editrice Ambrosiana, Milano 2000. In about one third of the twins that are born, the derivation from one single fertilised egg is recognised, and it is therefore defined *"monozygotic"*. This definition is demonstrated by molecular tests, which characterise the same genome of *monozygotic* twins and the sharing of 50% of the genetic features in *dizygotic* twins. In twins with two separate placentas (so-called *bichorial*) the separation into two parts of the original blastomeres of the inner cell mass took place before the fourth day following fertilisation, the moment in which the trophoblast (layer of tissue that is the precursor of the placenta) has already been defined. In the most common cases in which *monozygotic twins* share the same placenta (so-called *monochorionic)*, each of which is surrounded by its own *amnios* (*diamniotic*)*,* the separation takes place before the formation of the *amnios*, that is between the 3rd and 8th day following fertilisation. In this case the twins are defined as *monochorionic diamniotic*. Much more rarely, even though the twins are completely separate, they are surrounded by a single *amnios* insofar as the separation took place between the 8th and 12th day. Owing to this characteristic they are defined as *monoamniotic monochorionic* twins. In each of these combinations, the twins are separate from each other.

brain (meninges and superficial part of the brain) with trunk and distinct faces; *xylophagus* twins, that share a single sternum; *ischiopagus* twins, conjoined at the pelvis, genitals and anus.

Combinations of these defects are also possible (e.g. cephalo-thoracopagus or thoracopagus-omphalopagos). According to the type of conjunction it is technically possible or impossible to surgically separate the twins, who in many cases die a premature death.

There are also asymmetrical or unequal conjoined twins, incomplete twins 'attached' externally to a twin that can be defined complete: the '*vanishing* twin' (known also as 'foetal resorption'), is the foetus that belongs to a multiple pregnancy, which dies in utero and is partly or completely reabsorbed by the co-twin[7]; 'parasitic twins' are formed when one twin begins to develop in the womb, but the co-twin does not separate completely and a twin takes on the main development at the expense of the other.

Unlike conjoined twins, one of the co-twins ceases to develop during pregnancy and takes on features that vary between the presence of vestigial structures and the presence of an almost completely formed twin. The twin that develops only partially is defined as parasitic, rather than conjoined, insofar as it is only partially formed and its development is entirely dependent on the functions of the complete co-twin. The independent twin is also defined autosite[8].

Incidence

Conjoined twinning is a rare phenomenon and is also to be found in the animal world. In the literature very differing numbers are published, varying from 1 out of 200,000 *new born babies* (0.5 per 100.000) to 1 out of 2,800 (about 36 per 100,000), 72 times higher[9]. A reliable estimate is included between 1 and 2 cases per 100,000. The incidence is variable in different countries and periods of time, and in the different typologies. Assuming an incidence of 1 out of 50,000, an annual frequency of 50 cases per week in the world can be estimated; 11 cases a year in Italy. These latest estimates refer to *conceived* twins, including abortions too. The epidemiological studies are poor.

[7] In some cases this phenomennoin is classified as the twin embolisaiton syndrome. The twin that dies and is squashed by the co-twin is defined as "papyraceous foetus". The vanishing twin can die due to a defect of the embedding of the placenta, a defect in development that causes the disfunciton of a principle organ or due to the presence of a chromsomic abnormality incompatible with life. Often the vanishing twin manifests itself as a blind egg (membranes without an embryo, insofar as the embryo died in the first phases of development and was absorbed).

[8] H.J. Landy, S. Weiner, S.L. Corson,, F.R. Batzer, *The "Vanishing Twin": Ultrasonographic Assessment of Fetal Disappearance in the First Trimester",* in "Am. J. Obstet. Gynecol.", 1986, 155 (1), pp. 14-19; H.J. Landy, B.M. Nies, *The Vanishing Twin,* in "Multiple Pregnancy: Epidemiology, Gestation and Perinatal Outcome", L.G. Keith, E. Papiernik, D.K. Keith, B. Luke (eds.), The Parthenon Publishing Group, New York 1995; pp. 569-71; D. Pelega, A. Ferber, R. Orvieto, I. Bar-Hava, *Single Intrauterine Fetal Death (fetus papyraceus) Due to Uterine Trauma in a Twin Pregnancy,* in "European Journal of Obstetrics & Gynecology and Reproductive Biology", 1988, 80 (2), pp. 175-176; J.L. Grosfeld, D.S. Stepita, W.E. Nance, C.G. Palmer, *Fetus-in-fetu: an Usual Cause for Abdominal Mass in Infancy,* in "Ann. Surg.", 1974, 180 (1), pp. 80-84; C.E. Alpers, M.R. Harrison, *Fetus in Fetu Associated with an Undescended Testis,* in "Pediatr. Pathol.", 1985, 4 (1-2), pp. 37-46.

[9] O.M. Mutchinick et al., *Conjoined Twins: A Worldwide Collaborative Epidemiological Study of the International Clearinghouse for Birth Defects Surveillance and Research*, cit., p. 276.

The evident vagueness of the statistical data leads one to think that the epidemiological research is lacking and must be reinforced[10].

II. Prenatal diagnosis

The increasingly widespread use of prenatal diagnosis techniques (at least in the technologically advanced countries) increases the possibility of carrying out the early diagnosis of this pathology by means of ultrasound and, more accurately, also by magnetic resonance. In these cases the opinion of the consultant plays an extremely delicate role, in the measure in which he has to inform the couple of the present and foreseeably future conditions of the twins, the success of the operation after their birth and the long term consequences. Nevertheless, while it is possible to diagnose conjoined twinning, it is difficult to give a precise picture of the pathologies that might be associated with it. The diagnostic instruments – as refined as they may be – are often insufficient to express definitive opinions. For this reason the consultancy must be given by a multi-specialist team able to give information concerning the complexity of the condition diagnosed[11].

III. Ethical problems

Even though this is a relatively rare phenomenon, it deserves a proper bioethical investigation owing to the tragic nature and complexity of the correlated issues, which often call for evaluations and urgent decisions by the subjects involved, the parents and doctors in particular[12].

At the centre of the bioethical debate is the question of the *possible separation*. The progress in scientific knowledge and biomedical technologies (paediatric, reconstructive and transplant surgery) has made separation surgery possible which, until some decades ago, was impracticable, thus increasing the survival percentage and the quality of life of the twins[13]. Such new possibilities for surgery raise various ethical issues with regard to the subjects legitimated to decide, the relationship between the possible risks and benefits for the life and quality of life and to the time at which to intervene.

It must first of all be underlined that the case of conjoined twins with only one head, extra limbs or parts of organs is not taken into consideration (the operation should not present ethical problems even if carried out only for aesthetic reasons, since in fact one single head identifies one single person),

[10] There are four ethnic group categories in which there is a prevalence of the phenomenon: Anglo-Saxon/Caucasian, Chinese, Latin-American and Latin-European. According to statistics, it appears that conjoined twinning is statistically more frequent in the Latin-American ethnic group. A majority has been found in the female sex (thoracopagus twins) and in the male sex (parapagus twins and parasite twins). In the calculation of the incidence existing cases, diagnosed and aborted (by miscarriage or abortion) are not recorded.

[11] In this document the NCB does not set out to go into the specific bioethical issue in the connection between malformations disciovered during prenatal diagnoses and possible consequent choices to abort, but considers it right to stress how complex, controversial and – in the opinion of many – painful this question is. Reference is made to a previous document on this issue, *Prenatal diagnoses* (1992), and, in this opinion, in point III.1, the concluding bioethical guidelines.

[12] On the one hand the lack of of scientific and epidemiological studies is surprising, and on the other, the considerable amount of ethical studies is.

[13] L. Spitz, E. Kiely, *Success Rate for Surgery of Conjoined Twins*, in “Lancet”, 2000, 356, p. 1765.

but reference is essentially made (with the exception of the case of the parasitic twin) to two persons, who can be characterised by having two heads and only one body, to having two heads and two bodies more or less complete and a variable number of limbs.

The cases do not present problems where the conjoining is minimum (strips of skin or tissues) and the separation does not entail particular risks. The most complex cases are those in which the conjunction concerns extended parts of the body and the sharing of apparatus and vital organs, whereby the surgery for their separation seems indispensable to save the life of both the twins or of one of the two possibly to the detriment of the other or appears necessary to improve the quality of life, but implies a high risk with negative consequences that are difficult to avoid in relation to the delicate nature of the situation.

The ethical specificity of the problem concerns the peculiarity and uniqueness of the condition: conjoined twins are two distinct human subjects, insofar as each one is self-organised, but at the same time with organs and parts in common, whose communality can be necessary for their reciprocal survival. Their existence experiences not only a condition of physical connection, but often also of integration and reciprocal dependence (one cannot live without the other), even in the distinction of the individuality/personality. The phenomenon of conjoined twins constitutes a borderline case of the oneself/other relationship, in a sort of 'unitary duality': they are seen as distinct but at the same time appear a unitary entity that forces them to have an integral interdependence. Another peculiarity is constituted by the fact that in many cases it is not possible to ascertain which twin depends on the other (apart from the case of parasitic twins).

In bioethical reflection the debate is above all on whether the integral physical independence (or the possession of a separate body) is an indispensable requirement to be considered a subject/individual that must be always guaranteed its 'own' specific integrity or whether we find ourselves before a different integrity, that is unitary and double at the same time, which must be recognised and respected when there are no imminent threat to its life, in its 'own' particular nature. What is the best interest to pursue for conjoined twins? What is the importance of medical diagnosis, the parents' choice, the possible assent expressed by minors and how to configure the adults' assent? What are and must be the limits of medical intervention on the body in the respect of the person?

The NBC highlights that the dramatic nature and complexity of the condition opens up dilemmas in which it seems that any ethical response or practical solution to the cases presented in reality is problematic. Proof of this are those situations that – made known to the public opinion by the media – have raised and raise contrasting emotional reactions by society, the difficult of evaluation by the bioethicists and complex decisions by parents and doctors.

In this opinion the Committee discusses two situations often referred to in scientific and bioethical literature, considering it opportune to distinguish the position of newborn babies/minors from that of mature minors/adults.

1. Newborn babies/ minors

1.1. A first situation is that in which there is no immediate danger to the life of the twins in the present and foreseeably immediate future, while separation

by surgery, even though technically possible, is highly risky for the life of one or both twins[14].

Differing bioethical standpoints were expressed within the Committee which are reported below with their reasoning:

a. A first line of thought[15] considers that this choice to try and separate the twins is not ethically justified. In such circumstances in which surgery is not necessary, as there is no serious threat to the life of the twins nor a foreseeable worsening of the prognosis in the immediate future, the separation surgery is considered disproportionate owing to the high risks, as it could seriously compromise the health of the life and health of the newborn babies or of one of them to the detriment of the other.

This standpoint appeals to the value of the twins' life, maintaining that the conjunction must not be considered in itself as an unacceptable condition. The physical, psychic and social limits that conjoined twins can suffer are ascribable to that general broad condition of disability set out by the UN Convention on the rights of persons with disabilities[16]. And two persons with serious disabilities have an equal right to life and treatment independently of the condition of their bodies, separate or not.

It was stressed that the seriousness of life must never be confused with the seriousness of treatment. It is not ethically or juridically licit to begin disproportionate treatment to avoid a life with disability. In other words, the modalities of treatment of conjoined twins cannot be evaluated with bioethical criteria that are different from those that must be used to evaluate the forms of treatment used for individuals with disabilities, considering that they are minors and cannot express their will. As far as the doctor is concerned, the deontological principle must not be neglected according to which before a situation of disability like that of conjoined twins, as before any other illness, they must always have the primary objective of doing their utmost to safeguard life and health (present and foreseeable in the future), with the only limit of never having to resort to persistent therapy.

In these cases it must also be considered that waiting, if the situation is not life-threatening, finds a further justification in allowing the parents 'to gain time' so that they, with the help of the doctors, might better evaluate the clinical condition of their children and arrive at the best solutions for their health. For these reasons high-risk separation surgery is not considered ethically justified as it would result in configuring a form of 'experimental persistence'.

In the light of the legal system in force it must also be borne in mind that if there is no serious and imminent danger it is not justified to intervene, placing the life of the minor at high risk. Should the parents or doctors express their

[14] The word 'risk' is used in the general sense, even in the awareness that the evaluation of the likelihood of risk needs a precedent, which in many of the cases relative to conjoined twins is lacking.

[15] Standpoint shared by the members: Profs. Stefano Canestrari, Bruno Dallapiccola, Antonio Da Re, Lorenzo d'Avack, Maria Luisa Di Pietro, Riccardo Di Segni, Marianna Gensabella, Assuntina Morresi, Andrea Nicolussi, Laura Palazzani, Vittorio Possenti.

[16] UN, *Convention on the rights of persons with disabilities*, 2006, to which Italy adhered and which has been adopted in its legal system, as expressed also by the NBC in a motion unanimously approved on 27 June 2008, in which it called for "the rapid approval of the law of ratification and implementation of the Convention". In art. 1, par. 2: "Persons with disabilities include those who have long-term physical, mental, intellectual or sensory impairments which in interaction with various barriers may hinder their full and effective participation in society on an equal basis with others".

dissent, it is up to the judge supervising guardianship to decide on the best objective interest of the minors.

b. A second line of thought, departing from the same philosophical assumption of the recognition of the value of life and health, considers that such value can be thought of only in terms of identification and considers conjunction an unacceptable compromising of human identity. It considers that individuality is also an essential element of such values and must be guaranteed when a reasonable possibility of success of the separation surgery exists even at high risk, as long as it gives hopes of success although scant.

The motivations can be two, different from each other, even though in fact partly overlapping and operationally converging.

A first motivation is that of those[17] who, denying that the conjunction might be defined as a form of disability and even as a form of ' acceptable' disability, consider that however high the risk of the separation may be it is still proportionate to the pathology to be dealt with. In the measure in which the separation is not technically possible and in the absence of any threat to life, the right of the conjoined twins to live must be recognised.

According to others[18], separation surgery would be justified insofar as aimed at guaranteeing the twins or one of them a certain quality of life. Conjoined life is considered contrary to a fulfilled realisation of the person, in so much that it annuls autonomous individuality and constitutes a form of 'abnormality' or 'serious anomaly' going beyond the very context of disability. It would be a serious responsibility to condemn someone a priori to a difficult existence from every personal and relational point of view, when the possibility exists – even though slender – of guaranteeing 'normality'.

c. A third line of thought[19] instead considers that it is not possible to abstractly judge the licitness/illicitness of high-risk surgery, even in the presence of a life-threatening situation.

Not only can there be forms of conjunction that give rise to particularly serious disabilities, but as is highlighted below, very often these forms of twinning are accompanied by malformations of the vital organs which will worsen in time. In these cases it can be ethically acceptable for the parents and doctors involved, on the basis of a reasoned evaluation of the situation of the specific case, to operate immediately even if the surgery is extremely risky.

1.2. A second condition is the one in which, on the basis of an objective clinical evaluation supported by empirical data, the clinical certainty is shown of the imminent and serious threat to the life and health of both the twins. The abstention from surgery by the doctor will entail the death of both the twins or a foreseeable worsening of the prognosis, while the separation of the two twins can present two outcomes: the first to save both lives; the second to save at least one of the two.

It must be remembered that very often these forms of conjoined twinning are accompanied by very serious multiple malformations, affecting organs

[17] Standpoint shared by the members: Profs. Luisella Battaglia, Francesco D'Agostino.
[18] Standpoint shared by the members: Profs. Salvatore Amato, Silvio Garattini.
[19] Standpoint shared by the members: Profs. Laura Guidoni, Monica Toraldo di Francia, Grazia Zuffa.

and/or entire apparatus, like for example the heart and circulation system, whose functioning is compromised in such a way as to make it impossible for both the twins to continue to live united.

Surgery to save both lives can be actually presented only in the condition of conjunction in which the missing organs could be transplanted and the body parts reconstructed. This is a very difficult situation to realise from a medical and practical point of view difficult.

On the contrary the possibility is more frequent of an operation in which it is known beforehand that it is not possible to save both the twins, but that there is a fair chance of saving only one of the two, the 'strongest' or 'healthiest', or the one that has greater anatomo-physiological chances[20]. The arguable expression 'sacrificial separation' is often used. It is not therefore a separation owing to the need to eliminate the impediment to the physical independence of the two babies, but of surgery determined by a serious condition of their health, such as to jeopardise their lives.

In these cases the particular difficulties in which the decision-making process comes about must not be neglected or made light of. Before the immediacy and urgency of surgery a difficult psychological condition of all the subjects involved in the decision is presented, whether they be the parents or doctors. It must be added that any decision taken by the parents should be founded on their real understanding of adequate and correct information, which is at times difficult, since not always do the doctors have a clear idea of the prognosis in the circumstances characterising the twins' health. Nonetheless this cannot and must not exempt the doctors from giving the parents all the information necessary so that they can take part in the decision-making process in full awareness. It is clear that adequate information on the risks of possible surgery, pain treatment and the uncertainty of the prognosis must be intensified beforehand.

The Committee retains it opportune to call the attention to some of the varying opinions and reasoning more frequently expressed on this condition in bioethical literature: those that for the most part were examined during the hearings and debate held during the discussion on the opinion, even if not necessarily shared completely or in part.

a. A first line of thought considers[21] that the parents' choice would be ethically understandable and acceptable who, following adequate medical information and also in contrast with this, in the uncertainty of the outcome of possible separation surgery, decide to abstain from the operation, even though knowing that such choice will entail the death of both twins or of one of the two. This position can be motivated by many arguments: by reasons of a religious nature, the intention to make a choice not made at the moment of the prenatal diagnosis (or that is the choice to terminate the pregnancy), by the refusal to assume the responsibility and the moral burden of a choice should the surgery permit the survival or the possible survival of one twin only, by letting 'nature run its course', by the parents' will to avoid making the twins suffer owing to the operation and possible successive operations, bearing also in mind the fact that

[20] The hypothesis whereby only one can be saved and both have the same probability of survival is considered as a 'textbook case', not realised in practice.

[21] Standpoint shared by the members: Prof. Carlo Flamigni.

often a poor quality of life for both can be hypothesised with considerable healthcare costs.

b. Another line of thought[22] is in favour of surgery aimed at saving both twins, but is ethically against any surgery that a priori will not allow the saving of both twins[23]. It is in fact believed that the protection of the gift of life must be understood as an absolute prohibition to cause death and, therefore, the inevitable dramatic choice of one of the two twins is considered illegitimate. The surgical separation would moreover question the principle of equality according to which all subjects must be recognised the right to life regardless of their condition of health/illness.

c. A further line of thought considers that in the condition of an imminent and serious threat to the lives of both the twins and in the face of an appreciable and reasonable forecast of a life-saving outcome for one of the twins – on the basis of rigorous clinical tests – the surgical separation must be considered ethically correct and the doctors' intervention justified.

The members of the Committee who shared this standpoint[24] were nonetheless keen to stress the fact that they do not share some of the arguments in literature supporting this thesis. To summarise: a) surgical separation is justifiable insofar as the 'weak' twin can be considered as a 'donor of organs', in the viewpoint of a 'self-sacrifice' for the wellbeing of the other with greater chances for the future; b) the death of the 'weak' twin is justifiable insofar as, according to a current terminology in literature, the 'unjust aggressor'[25], who threatens the life of the strong twin who, before such threat, can legitimately defend itself (in other words, a reaction against the aggressor would be justified even if it causes his/her killing in so much as being the only means to save his/her own life); c) the 'weak' twin is 'destined to die', would therefore die anyway, and thus it is allowed to make him/her die in advance; d) surgery is justifiable with reference to the theory of the 'double effect': the realisation of an unnecessary planned operation to obtain the survival of only one baby (intentional positive effect) produces the death of the 'less healthy' child as an unwanted negative effect.

The criticisms of these arguments can be summarised as follows: a) in this context the donation of organs cannot be invoked insofar as it presupposes the informed consent and the absence of risk for the life of the donor: the common shared organs – if equally connected to the physiological systems of the twins – are part of the corporeity of both and cannot be arbitrarily taken away from one to the advantage of the other; b) the 'weak' twin has no capacity to act and therefore to attack; the lack of will and intentionality determines the failure of any form of charge and can instead represent an involuntary 'biological threat'

[22] Standpoint shared by the members: Prof. Maria Luisa Di Pietro.

[23] This is what happened in the case of the Maltese parents with their conjoined twins Jodie and Mary, a widely debated case in bioethical literature.

[24] Standpoint shared by the members: Profs. Salvatore Amato, Luisella Battaglia, Stefano Canestrari, Bruno Dallapiccola, Antonio Da Re, Francesco D'Agostino, Lorenzo d'Avack, Riccardo Di Segni, Silvio Garattini, Marianna Gensabella, Laura Guidoni, Assuntina Morresi, Andrea Nicolussi, Laura Palazzani, Monica Toraldo di Francia, Giancarlo Umani Ronchi, Grazia Zuffa.

[25] Terminology taken from the debate on the unjust war and legitimate defence since Scholasticism, but already present in the Talmudic debate that speaks of the right of the rodèf, "the pursuer".

for the life of the other; for some therefore it cannot be considered an ‘unjust aggressor’, for others it is nevertheless an aggressor, even though an unintentional one; c) the ‘destination to death’, as foreseeably certain as it may be, does not amount to death; such future event is not sufficient to justify the decision to make a subject die in advance to save the other; d) the ‘double effect’ theory – often used also in the context of medical ethics – foresees different versions, formulations and expressions referring to different and even contrasting theories and assumptions, often giving rise to equivocal ones.

In such context the surgical separation is instead justified for these members of the Committee on the ethical and juridical principle of the protection of life. Therefore, the emphasis is placed primarily on the hope of saving both twins, but if this is not possible, at least of guaranteeing one of the twins a prospect of life that would be inevitably precluded by the choice to let nature take its course.

The aim of the operation is not to bring about death but to save a life and it is justified in the perspective in which there are no other alternatives and it is not possible to operate for the benefit of one without negative consequences for the other, who must nevertheless receive all the medical attention necessary to guarantee a painless end.

It is not therefore a question of choosing the ‘strongest’ of the two twins ‘at the drawing board’, or of evaluating that the life of one person has more or less value (against the principle of equality), but of the ascertaining the medical response by the parents, on the basis of rigorous clinical tests, according to internationally recognised scientific standards, according to which it can be excluded that one of the two twins has any, even though slender, chance of living and that the continuation of his/her existence determines the death of the other twin that could otherwise be saved. The option to ‘let nature take its course’ in the medical certainty of the death of both, would represent negligent conduct and the abandoning of therapy as it would prevent the saving of the saveable life.

In all these cases the surgery must be ‘reasonable’, or that is, it must exclude those levels of extreme experimentation whereby the suffering of the persons subject to it – newborn babies in this case – is not balanced by adequate life expectations.

2. Mature minors and adults

The decision to surgically intervene or not, both in conditions of necessity and non medical necessity, takes on a different moral relevance if the conjoined twins (extremely rare cases) are adult and both competent, or if they are mature minors. In this case the informed autonomous consent/dissent of the subject’s decision in the request must be recognised, with prior exhaustive and calibrated mandatory consultancy with respect to the conditions of the subjects by the doctor or even better a medical team.

Such expression of autonomy raises a number of moral problems in consideration of the peculiar condition of the conjunction of corporeity.

In the situation in which both wills are in agreement with respect to operate or not operate in the consideration of the possible future scenarios outlined by the doctor, also in the measure in which the surgery or non-surgery can

inevitably bring about the death or threaten the life of one of the twins, such decision is generally considered acceptable (if the operation is proportionate).

Cases of conflict are not known of, insofar as conjoined twins have until now expressed agreement both in their acceptance and refusal of treatment. Their life of being always conjoined brings about not only a physical but also psychic and emotional union (even though having different personalities), to the point that the adult twins – in the measure in which they are competent – positively assess their life and their peculiar conditions; they refuse to be separated even if it is the only condition to continue to live; they refuse to be separated in the case in which they know that their survival would entail the death of the other twin[26].

In the measure in which – abstractly – a possible contrast of will takes shape with respect to the choice of separation, given the risks of the operation for the subject that chooses, but at the same time for the subject that undergoes this choice even though not sharing it, it must be considered that it is not only the individual right to self-determination that is being questioned, with all the problems relative to the decisions that can jeopardise life, but also the duty that the twins have to respect each other's integrity. For this reason therefore the choice of the twin refusing treatment must be privileged so as not to jeopardise his/her own life.

Another extreme possibility is represented by the choice of one of the twins to sacrifice him/herself for the other. The details of the case record will not be dealt with in this opinion but only a general ethical criterion will be formulated: the assent of the patient never represents an obligation for the surgeon to operate, while the dissent represents an obligation to not operate. There is then the absolute right of the mature minor to be informed for a consent or conscious dissent, since also before complex existential choices adolescents manage to prefigure the future and to assume the responsibility in conformity with their own life project[27]. Choices that confirm the inappropriateness to establish rigid criteria that establish the acquisition of the full capacity to act and how, on the other hand, a case by case evaluation is opportune of the capacity of each single minor finding him/herself in that specific situation.

IV. Bioethical guidelines

The reflection of the NBC has highlighted the problematic nature of the evaluation of the question of the treatment of newborn/minor conjoined twins, both by parents and doctors, in relation to the choice to intervene or not to separate them in the various clinical cases outlined above. All the more so that such standpoints, expressed theoretically (in order to give guidelines and direction in particularly difficult cases at clinical and ethical level), must always and nonetheless be pondered on the basis of relevant objective data and rational reasoning, in the context of the complexity and variability of the actual cases.

[26] In the rare cases known until now, many conjoined twins refused to be separated upon reaching maturity. For example, in 1967 Mary and Margaret Gibb refused separation, even when the imminent death of one of them owing to a tumor would have meant the unavoidable death of the other.

[27] Principle which appears in various regional and international documents, among which: *Convention on the rights of the child* (UN, 1989, art.12) and *Convention on the human rights and biomedicine* (Oviedo, 1997, arts. 6, paras. 2 and 24).

Despite the above mentioned divergences, the Committee arrived at the formulation of a number of shared recommendations that may constitute a conceptual reference horizon to foster ethically complex decisions at clinical level:

a) research on the causes of the phenomenon of conjoined twinning must be promoted and increased, at scientific and epidemiological level in order to understand its etiology and pathogenesis. Conjoined twins cannot only be considered a problem to be avoided (by prenatal diagnosis), but also to understand, prevent and treat with an interdisciplinary approach. The professional training of doctors and healthcare staff must furthermore be increased so as to build a team able to deal with such complex cases, also in suitable healthcare facilities;

b) the information to the parents, within the prenatal diagnosis of conjoined twinning – also owing to the limits of the technologies that can be used in this phase to define the anatomy and physiology of the origins with precision – must be given by a multi-specialist team of doctors, made up of different consultants in relation to the typology of the conjunction, or that is to say, with a close examination of the possible implications in relation to the chance of living, the quality of postnatal life, the success of the separation by surgery. It must be remembered that the information is not always clear and definitive, given the complexity and the unforeseeabilty of the condition. The information must be continuative – for the duration of the entire and complex course of treatment – according to the evolving of the conditions and must bear in mind the ability to understand and psychological-emotive difficulty of the parents, envisaging also help and psychological support in therapeutic alliance;

c) there must be no surgical intervention in the cases in which clinical conditions of persistent or experimental therapy are presented. Even though recognising, owing to the exceptional nature and complexity of the cases, the difficulty in establishing a clear demarcation line between proportionate and disproportionate or experimental interventions/treatment, it is to be hoped that in the various cases the parents and doctors can come to unanimous choices in the search for a balance between the prospects of life, quality of life and therapeutic needs;

d) considerations relative to the costs of the healthcare assistance must not come into the ethical evaluation of the duty to operate or the abstention from surgery;

e) conjoined twins have the right to rehabilitation, the continuity of treatment and integrated physical-psychic-social assistance;

f) the role of the ethics Committee is important in supporting the decisions to be taken by parents and doctors;

g) the possible clause of conscience raised by the doctor or the medical staff for the assistance that is in conflict with their clinical and moral convictions, must be commensurate with the particular nature of the intervention and the

circumstances, especially in the case in which the professional, with his skills, represents a ‘crucial’ and irreplaceable element for therapeutic success;

h) the confidentiality of the case must be respected, even to prevent the mass media from disturbing the delicate decision-making process;

i) in the case of adults, the will of the twins must be considered identical to that of any other competent adult, even with reference to the choice to undergo experimental treatment or to refuse therapy.