Anaesthesia of conjoined twins

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Abstract

Conjoined twins have been a source of fascination to the public and the medical profession for centuries. Their birth was initially viewed as an ominous sign of impending disaster. Since Middle Ages into the 19th century they were regarded as monstrosities and were exhibited at circuses and sideshows. The frequency of conjoined twins is approximately 1 in 50,000 gestation, but many of them die in utero, are terminated or stillborn. The true incidence is estimated to be 1 in 200,000 live births. This article gives an overview of Siamese twins and of the prenatal diagnosis in assessing the prognosis, anaesthetic and post-natal surgical management and outcome. Anaesthesia for conjoined twins surgery, whether prior to or for separation, is an enormous challenge to the anaesthesiologist. The site and complexity of the conjunction affect management of the airway, an intravenous access, the extent of blood and number of surgical specialties involved. Preoperative assessment and planning with inter-disciplinary communication and cooperation is vital to the success of the operations. Meticulous attention to detail, monitoring and vigilance are mandatory.

Key words: conjoined twins, anaesthesia; conjoined twins, diagnosis; conjoined twins, management

Siamese twins — conjoined twins, are not only an extremely rare congenital malformation in humans but also the most difficult one to treat. The defect is an example of one of the most complex organizational and ethical issues encountered in medicine. According to different epidemiological reports, the incidence of conjoined twins is estimated to range from 1 in 50,000 to 1 in 200,000 births, depending on geographical region. The majority of Siamese twins are stillborn or die shortly after birth due to accompanying developmental defects, hence they constitute an extremely small group of patients. The defect is three times more frequent in girls [1–6]. Its occurrence has been found to vary significantly by geographical region and race. It is the least common in Caucasians; in India and Africa its incidence is markedly higher than in Europe or America [7, 8].

Currently, the most probable theory explaining the formation of conjoined twins is that of incomplete separation of two embryos between the 14th and the 18th day following fertilization [2, 9, 10]. It is believed that the environmental and maternal factors that result in monozygotic monoamniotic pregnancy can also be the cause underlying the formation of conjoined twins, which is supported by clinical observations. Conjoined twins are always of the same gender and genetically identical. They are always joined with the same body parts and are positioned symmetrically [8, 11, 12].

The most clinically useful, albeit highly simplified, classification of conjoined twins divides them into symmetrically conjoined, i.e. of the same size, symmetrical to each other and asymmetrically conjoined, i.e. one is always smaller (a parasite), and dependant on the other [13].

Symmetrically conjoined twins include children joined at the following regions:

A. chests (thoracopagus, xiphopagus) with possible fusion involving the heart, liver and upper gastrointestinal tract, or abdominal cavity (omphalopagus) — united at the region of the liver and gastrointestinal tract where children are facing each other,

B. sacral bones (pygopagus) — children with their backs to each other, usually joined at the nervous system, the crotch and anus,
C. heads (craniopagus) — usually fused medially sharing the nervous tissues, large vessels and sinuses,
D. pelvis (ischiopagus) — most frequently sharing the urogenital system, rectum and liver. Children can have either 4 fully developed lower limbs or only 3 with 1 deformed; this defect may be also accompanied by omphalocoele [6, 13]. The group of asymmetrical conjoined twins includes cases in which the body parts differ in size — one foetus, called a parasite, is smaller, partially developed and dependant on the other, fully developed foetus, called an autosite (Fig. 1) [11, 13].

Advances in paediatric anaesthesiology and intensive care facilitated the development of paediatric surgery enabling management of congenital malformations in newborns and infants, including the complex ones, such as Siamese twins. Each case of conjoined twins creates an unusual situation in which the attending team has to take an appropriate decision about possible treatment. Siamese twins, although having one organism, are two individual persons [14, 15].

Thanks to the popularization of non-invasive diagnostic procedures in modern obstetrics, such developmental defects can be early detected, parents prepared psychologically and labours planned near the centres where the defects can be fully diagnosed and separation surgery performed [13, 16–18]. The above strategy, applied also to other malformations, gives better chances for favourable treatment outcomes. It eliminates iatrogenic effects of prolonged transportation on the child’s health condition and enables proper preparation of the specialist centre [5]. It should be however borne in mind that Siamese twins are such a complicated developmental defect that even the use of the most advanced diagnostic methods does not always allow to assess accurately the configuration of twins’ internal organs, hence final verification is possible only on the operating table [19]. Siamese twins are more difficult to transport than single newborns; therefore, the transport team should be suitably prepared and equipped. It is essential to try to stabilize the child’s condition prior to transportation, particularly to secure airways and respiratory efficiency, maintain proper positions during transportation, ensure normothermia and provide adequate fluid supply [20]. Cardiopulmonary resuscitation, should it be necessary, is very difficult and highly risky, especially in children joined at the thoracic and abdominal region. Due to defective anatomy, chest compressions may not be possible and cause damage to the intestines and liver.

Spontaneous labours in cases of undiagnosed pregnancies with conjoined twins usually result in severe injuries to at least one of the twins [6, 13]. Perinatal injuries endanger the health and life of a mother and her children and force emergency separation, which always increases the risk of treatment failure [6, 21].

The majority of authors experienced in the treatment of this malformation unanimously stresses that separation surgery of Siamese twins should not be performed during the neonatal period. However, it should always be taken into account that the condition of one of the twins can deteriorate and emergency operation can be necessary [6, 12, 13, 19, 21]. It has been demonstrated that the postponement of separation surgery until the twins are several months old, unless life threatening, increases the chances of good outcomes. We should not however wait too long due to increasing problems, both physiological and psychological [2, 22].

The basic principle of surgical treatment of conjoined twins is a proportional division of organs enabling children to live separately; a favourable treatment outcome requires adequate amounts of tissues and organs necessary for the two beings to function independently. In cases where children cannot be separated due to the type of conjunction, treatment and care should be provided until therapeutic options are exhausted. Under no circumstances should the decision of treatment abandonment be taken. Separation of Siamese twins, where division of all organs allowing independent life is possible, seems expedient and permissible, even if it means severe disability. All decisions concerning surgical treatment of conjoined twins should be taken considering the fact that we deal with two independent human beings and cannot decide that one life is more valuable than the other [23].

The general health condition of conjoined twins without perioperative injuries and additional defects is usually good during the first days following birth and their treatment focuses on appropriate care and nutrition. Siamese twins planned to be separated almost always require prior, often repeated, general anaesthesia or sedation for diagnostic examinations, for treatment of problems resulting from accompanying defects of the gastrointestinal and urinary tract or for insertion of central venous catheters and expanders. In some cases (e.g. the shared part is damaged—usually the liver, circulatory failure increases or sepsis develops), it is necessary to provide anaesthesia for emergency surgery [2, 6, 19, 22].

Adequate and detailed planning as well as repeated reviews of surgical treatment by all the specialists involved, who have at their disposal full diagnosis of the malformation, are of extreme importance for both the surgical and the anaesthetic team. Only such proceedings allow correct planning of the anaesthetic procedure at each stage of surgical treatment of conjoined twins.

The anaesthetic management prior to separation of Siamese twins is planned based on current information obtained with successive diagnostic tests and has to be
Figure 1. Classification of conjoined twins [drawing Elżbieta Gazda]

Rys. 1) Thoracopagus
Rys. 2) Craniopagus
Rys. 3) Cephalothoracopagus
Rys. 4) Ischiopagus
Rys. 2) Xiphopagus
Rys. 2) Pygopagus
Rys. 7) Dicephalus
Rys. 8) Heteropagus (acephalus acardius), asymmetric form of conjoined twins
modified depending on new information on organ connections. It allows to assess the extent of shared circulation and to plan the type and sequence of anaesthesia induction as well as to select drugs to be used to induce and maintain general anaesthesia. In cases of Siamese twins joined at chests, abdomens or heads, shared circulatory systems of both children should be expected, which is essential for induction of general anaesthesia, especially intravenous one [21–24]. Furthermore, shared circulatory systems can lead to their uneven development after birth due to differences in metabolism of conjoined twins [25].

Participation in the assessment of conjoined twins based on the results of successive diagnostic tests as well as care during their stay at the intensive care unit (ICU) allow to estimate potential difficulties likely to occur during intubation, especially in twins joined at chests. Emergency intubation is always extremely difficult; therefore, it is necessary to monitor carefully children to be able to design the procedure in advance. In the majority of cases, the use of a laryngeal mask is not recommended.

In cases of twins conjoined at chests and abdomens, the fact that twins are facing each other hinders the maintenance of patent upper airways during the induction of general anaesthesia with a face mask. Moreover, it is difficult to expose the larynx during intubation. The rotation of children's heads from the lateral position to close to the sagittal one in order to facilitate intubation may in turn deform the upper airway [21, 26–29]. Some authors are of opinion that prolonged conjoined twinning may lead to the development of severe cervical lordosis and inhibition of mandible growth, which additionally increases the difficulties in intubation [30–32]. After intubation, it is essential to fix the endotracheal tube. Frequent changes in position of children during surgery pose a risk of dislocation, or even accidental removal of the endotracheal tube, which may lead to intraoperative threat to the twins' lives [33]. The best measure to protect the endotracheal tube from dislocation or accidental extubation is intubation through the nose and correct taping of the tube followed by continuous monitoring.

The plan of anaesthesiological management should take into account the type and route of premedication, drugs to be used to induce and maintain anaesthesia, as well as the extent and type of monitoring of vital signs during general anaesthesia.

Anaesthesias for diagnostic examinations and pre-separation procedures require a different range of monitoring – only the non-invasive one. While selecting the anaesthetic method it is always a priority to be able to wake the children immediately after the diagnostic or pre-separation procedure and refer those with normal cardiopulmonary function to the ICU [28]. Separation surgery requires different anaesthesiological management. Preparation of conjoined twins for anaesthesia prior to separation surgery, which involves the induction of general anaesthesia, intubation, provision of peripheral and central venous accesses, direct arterial access, appropriate positioning of patients on the operating table and protection against heat loss, is time-consuming and can take even several hours [10, 29].

Anaesthesia for diagnostic and final separation procedures is always conducted by two independent anaesthesiological teams; the operating theatre should be equipped with two breathing machines with full monitoring. Before the initiation of anaesthesia we have to decide on which twin is going to be anaesthetized first, on the method of induction of anaesthesia and intubation. The preferred method is sevoflurane inhalational induction. In twins where no difficulties in airway maintenance are anticipated, intravenous induction can be used. Anaesthesia is maintained by combining inhalational anaesthesia and fractionated doses of opioids or applying intravenous anaesthesia, in accordance with preferences and experience of the anaesthetists performing the procedure. Relaxation of striated muscles is achieved with drugs most commonly used in paediatric anaesthesia (rocuronium, vecuronium, atracurium). In the majority of cases, the separation procedure lasts several hours, and therefore pancuronium is preferable. Drugs for perioperative antibiotic prophylaxis are selected according to hospital recommendations.

In order to increase the safety of conjoined twins during anaesthesia and separation surgery, all measurement and infusion lines as well as respiratory systems are coded with two colours (different for each child), which makes it easier to distinguish the twins during the surgery and to avoid mistakes. A vast majority of separation surgeries requires several changes of twins' positions on the operating table. Therefore, it is necessary to remain extremely vigilant and careful when maintaining the patency of airways. Meticulous care for appropriate positions on the operating table, with the use of various protective rests securing the limbs and releasing diaphragms for proper intraoperative ventilation, is of prime importance.

In paediatric anaesthesia, strict dosing of drugs takes into account the precise body weight of a child. During anaesthesia of conjoined twins each child is given half of the dose of drugs and fluids calculated for their join body weight. After separation the dosing schedule changes assuming that the body weight of each child is half of their join weight before separation. Once the children are admitted at the ICU, they are weighted and subsequent dosages of drugs and fluids are calculated at their actual body weight.

Estimation of intraoperative blood loss in conjoined twins is extremely difficult. Sometimes bleeding from the
common surgical wound is different in each twin. Blood loss is estimated on the basis of measurements of blood volume sucked from the surgical site to the suction container, amount and weight of the dressing material used, values of haemodynamic parameters, haematocrit and haemoglobin determined at constant intervals. Half of the estimated volume of the blood lost is transfused to each child [2, 6, 9, 26].

Furthermore, various problems can also be associated with too many members of the personnel present at the operating theatre, which is a result of the interest this unusual surgery evokes. Therefore, some authors prefer to perform these surgeries on days off, not to disturb normal functioning of the operating suite and to avoid many observers at the operating theatre [21, 29–31]. It is also recommended to carry rehearsals at the operating theatre, which allow determining the exact role of each specialist involved in separation surgery [2, 34].

Another serious issue during separation surgery is hypothermia caused by the extensive surgical wound, which increases heat loss by evaporation, radiation and convection. Therefore, it is necessary to use all the available methods to protect children against heat loss at the operating theatre. Many authors emphasize that preservation of normothermia during separation surgery is one of the most important factors affecting the outcome of surgical treatment [21, 22, 32, 35].

Separation of conjoined twins can last even several dozen hours; therefore, full-range monitoring of patients is necessary and invasive monitoring is a rule. Laboratory tests during surgery, often every 1–2 h, should also be planned.

Conjoined twins, who cannot undergo separation due to the defect configuration, are treated and cared for at the ICU according to the rules that apply in such situations. The anaesthesiologist attending children at the operating theatre and ICU encounters many clinical situations that create moral dilemmas. Thanks to the progress in intensive care, children of considerable degree of prematurity, with various genetic disorders, serious malformations that prevent independent life can be treated, which often raises certain ethical concerns among the treatment team. Despite long-term expensive treatment, many of these children cannot be saved and on many occasions, their treatment leads to severe, irreversible complications [36]. In cases of Siamese twins with conjunction that does not guarantee the survival of both of them (for example thoracopagus with the common heart), it is possible to distinguish at least two opposing points of view of ethicists, lawyers and doctors. Such cases provoke a fundamental question: is the decision of sacrificing one life to save another ethically justified?

None of the youngest patients is able to give informed consent to the treatment suggested or to refuse it, assuming at least part of moral responsibility for therapeutic decisions. Parents of these children are required to help doctors with therapeutic decisions in their own interest.

In recent years, owing to the media, we were able to witness doctors’ decisions concerning the separation of one of the rarest types of conjoined twins, i.e. twins connected with their heads. In 2001, the team of doctors from Singapore supervised by Goh, after the surgery lasting 97 hours, managed to separate 11-month-old Nepali girls. In July 2003, the same team of neurosurgeons attempted the separation of 29-year-old Iranian sisters — Laleh and Ladan Bijani — also joined at their heads. The sisters made a conscious decision of separation even though the doctors estimated the chances of success at 50%; one wanted to be a journalist, the other a lawyer. They both died during surgery. Their independent, certainly well-thought decision, divided doctors and ethicists [37]. Separation surgery of conjoined twins is not always necessary. American Siamese sisters, Lori and Reba Schappell, conjoined by the heads, who were born over 40 years ago in Pennsylvania, prove it possible to live a life without separation. Despite being connected by the frontal lobe, Lori and Reba have different inclinations and habits. Although inseparable, the twins differ in personalities. They lead an active life — Rega is a country singer, and Lori runs their home.

Currently, there are at least several pairs of conjoined twins in the world that are socially adapted and lead almost normal lives, despite not being separated. Thus, the question whether the separation of conjoined twins is always necessary, regardless of circumstances and risks involved, remains an unanswered ethical dilemma.

References:

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