

Intensive care of conjoined twins

Krzysztof Kobylarz

Chair of Anaesthesiology and Intensive Therapy, Department of Anaesthesiology and Intensive Therapy, University Paediatric Hospital, Collegium Medicum, Jagiellonian University, Kraków, Poland

Abstract

Conjoined twinning is one of the most uncommon congenital anomalies. Maintenance in an intensive care setting during this time allows for close monitoring, stabilisation, and nutritional supplementation of the infants as necessary to optimise preoperative growth and development. The birth of conjoined twins is a very difficult and dramatic moment for parents. It is also a very difficult situation for the team of physicians, nurses and other required hospital staff to carry out treatment and care of these specific developmental anomalies. The diagnostics and treatment in this extraordinary situation requires close cooperation of the multidisciplinary medical team, which includes their personal experience and medical knowledge, with a team of intensive care unit nurses. This report presents the rules in case of conjoined twins during their intensive care unit stay with special reference to the proceedings before and after complete separation.

Key words: conjoined twins, intensive care; conjoined twins, diagnostics; conjoined twins, management

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Conjoined twinning represents one of the rarest human congenital malformations. Most conjoined twin cases require intensive care unit (ICU) treatment. The ICU management of children with this malformation is unique and challenging for physicians, nurses and other hospital personnel with regard to patient diagnoses, treatments, care and rehabilitation.

An appropriate communication system that enables strict cooperation of the highly specialised physician and ICU nursing staff team is required to care for, diagnose and treat conjoined twins. Thanks to this system, difficult therapeutic decisions can be made promptly, if need be. Experienced nurses that care for the conjoined twins and are continuously in contact with and are observing them are essential for proper information flow amongst the various physician teams involved in patient management [1].

The conjoined twin birth is an extremely difficult and dramatic moment for parents. Notwithstanding, the early prenatal diagnostic procedures and information about malformations that is provided before delivery makes the first contact with the conjoined twins an extremely stressful event for their parents and evokes fears of their fate. It is vital to provide them with accurate information about the treatment options and their consequences, which often involve

severe disabilities [2, 3]. Additionally, the parents should be allowed to adapt to the ICU setting, which is initially unusual for non-professionals.

In the initial ICU hospitalisation period, conjoined twins are to be separated, and for those in whom separation is infeasible because of present treatment- and care-related difficulties and problems, basic nursing and diagnostic procedures are carried out first. In life-threatening cases and when the anatomical configuration of the malformation enables separation, the diagnostic procedures are usually confined to blood tests, other basic laboratory tests, plain films of the entire bodies of the children and ultrasound examinations, as a result of time limitations [4–8].

Special efforts by the nursing staff are needed while taking care of Siamese twins, especially prior to surgical separation. The main concern is to provide appropriate care for these unusual patients. Specifically, these patients are unusual because although they are two united children, they have two separate lives have to be cared for simultaneously, which is extremely challenging for the attending team and requires special nursing and therapeutic-rehabilitative procedures (Figs 1–4) [3–5].

Conjoined twins require continuous vital sign monitoring with both monitoring devices and thorough observa-



Figure 1. Conjoined twin nursing procedures are always provided by two nurses



Figure 2. Conjoined twin care in the ICU setting



Figure 3. Conjoined twin bathing in the ICU setting

tions by the attending nursing team for many weeks to months [2–4]. Two nurses should perform all of the monitoring procedures, with each caring for one of the twins. Even



Figure 4. ICU rehabilitation procedures

during simple activities, such as feeding, nurses should consider and remember that simultaneous filling of both stomachs in *thoraco-omphalopagus* twins is inadvisable as abdominal distension can cause respiratory difficulties in one or both twins. The twins should be fed frequently and in turns to provide the calorie requirements for both and to avoid the respiratory disorders caused by distention. Feeding requires patience (as such twins eat throughout the day) with short breaks for sleep, especially at the beginning of their extra-uterine life. After they have received several dozen millimetres of food, they do not want or cannot store more food and have to rest for several dozen minutes before eating another small portion. Additionally, proper positioning of *thoraco-omphalopagus* twins for feeding is extremely difficult.

Adequate fluid and calorie supply provisions are a precondition for the proper development of both twins. To prevent dehydration or overhydration, the degree of hydration should be regularly assessed clinically (fluid balance, daily measurements of body weight, stretching of the fontanel, subcutaneous area filling) and biochemical parameters should be controlled. When any disorders are found, they have to be corrected promptly.

In co-existing gastrointestinal defect cases and when colostomies are necessary, the risks of urinary infections and abdominal skin inflammatory lesions are higher, especially when accompanied by diarrhoea. When the gastrointestinal tract cannot be used to meet fluid and calorie require-



Figure 5. Nursing issues in conjoined twins

ments, parenteral feeding should be introduced, which creates further treatment and care difficulties. The access to central vessels and their proper management requires much more effort in conjoined twins than in a single child, which is associated with the anatomy of the conjunction. In conjoined twins that are to be separated, the surgery and stable central venous access maintenance should always be taken into consideration. Each vascular access should be “spared” as their numbers are limited in these children [7, 9]. It is necessary to provide additional access protection against their accidental removal during nursing procedures or by the twins in later developmental periods. When the twins have aged several months, one twin can accidentally remove the jugular or subclavian vein vascular accesses of the other child, particularly in the upper part of the body. There are cases in which children “fighting” with each other or pushing each other caused scratched faces. The separation of twins with a soft toy can be helpful (Fig. 5).

During routine nursing procedures, such as bathing, the cannula can be easily and unintentionally removed. Femoral vein catheters can be better protected against such removals, yet appropriate regimens that protect against infections in this region are more difficult, especially in intestinal fistula cases that are associated with co-existing gastrointestinal defects. After several months in the ICU, peripheral vessel accesses in the limbs are usually exhausted. When difficulties in providing stable central vascular accesses are anticipated, tunneled catheters or vascular ports should be considered [9].

An additional difficulty, in which ICU anaesthesiologists that attend conjoined twins often face, is maintaining respiratory efficiency. Siamese twins, even during respiratory efficiency periods, present a special example of the respiratory mechanics, particularly *thoraco-omphalo-ischiopagus* twins. In this conjunction type, the bony thoracic cavity

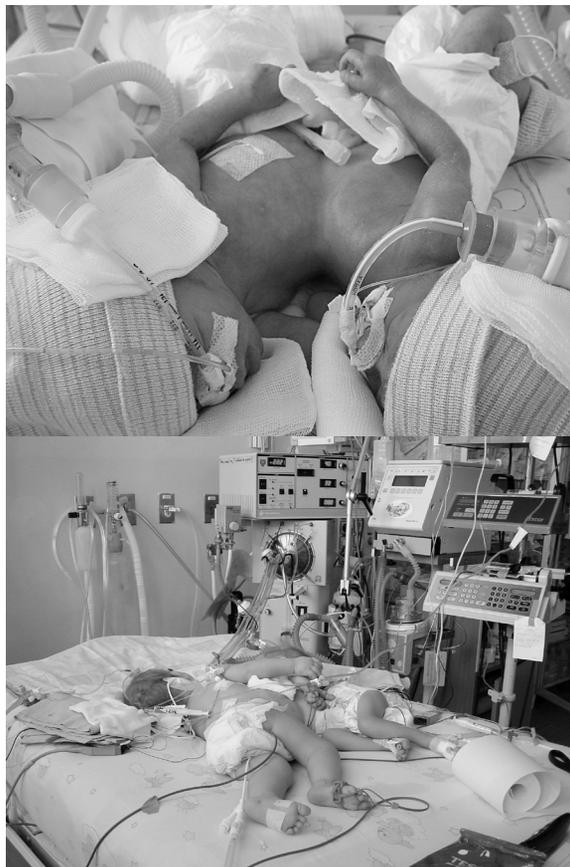


Figure 6. Respiratory failure management in conjoined twins

scaffolds are smaller or joined to a considerable degree, and both diaphragms can also be united. Two independent respiratory centres stimulating the patients’ respiratory muscles generated two different and independent respiratory rhythms. In children whose chests are united to a higher extent, paradoxical movements are observed. Due to the lack of synchronic respiration, children start to “quarrel” (especially during agitation), which leads to deteriorated gas exchange, which is confirmed by reduced SpO_2 values in both twins. In children sharing chests and abdomens, a difference in spontaneous respiratory efficiency has been observed, depending on the side their bodies are placed. Likewise, bronchial tree clearance of physiological secretions is more effective with a synchronous cough reflex. In developing respiratory failure cases, it is extremely difficult to determine indications therapy when a ventilator is being used. The biochemical, radiologic and clinical criteria used in other children are not sufficient. In conjoined twins, the physician’s experience is of utmost importance, and based on clinical increasing respiratory disturbance assessments and all of the laboratory test results, the physician should determine the degree of risk and initiate conventional ventilation therapy in advance [10, 11] (Fig. 6).

Frequent positioning onto the patients' side (due to the anatomy of conjunction) results in incomplete expansion of the dependent parts of lungs, which predisposes them to the development of atelectasis and inflammatory lung conditions. Therefore, proper care and rehabilitation provisions that consider all of the needs of the developing children are essential. When endotracheal intubation and respiratory support are decided, much bigger intubation problems should be anticipated in conjoined twins than in unaffected children [10–15]. In cases when respiratory efficiencies are deteriorating and quick intubations are required, the intubation times in conjoined twins is longer. Additionally, intubations are much more difficult when one physician is on duty without the assistance of another physician to help carry out the procedure. The above should be considered when planning duties. When respiratory failure develops in both children, intubation ought to be started in the child whose respiratory efficiency is worse. Another difficulty is the proper (i.e., stable) endotracheal tube fixation in the first of the conjoined twins to prevent a spontaneous extubation during position changes when intubating the other child [7]. To assess whether the endotracheal tubes are properly placed, respiratory murmur auscultations may not always be sufficient; therefore, chest X-rays are required. While using supportive or substitutive therapy due to respiratory failure in one or both twins, the current recommendations concerning mechanical ventilation should be followed. Since the 1990s, pressure-controlled ventilation (PCV) with respiratory volumes up to 10 mL kg^{-1} is most commonly used. Higher volumes, even 15 mL kg^{-1} , were used prior to this time. To determine the respiratory volume for each child, the actual body weight of one child and not both children should be taken into account.

Respiratory failure in conjoined twins that lasts longer than 6–8 weeks is a tracheostomy indication. Its advantage is the possible reduction or even elimination of pharmacological sedation because children better tolerate tracheostomy tubes than endotracheal tubes. Moreover, tracheostomy enables for oral feeding, weaning from a ventilator and spontaneous respiration [11, 15].

My experiences show that in increasing respiratory failure cases, early intubation is beneficial, and thanks to early intubations, hypoxia in one of the twins can be avoided. In prolonged respiratory failure cases, an early decision to perform tracheotomy helps to solve the problem, especially in *thoraco-omphalopagus* twins.

After the placement of expanders (a silicon balloon connected with a special entry that enables for multiple fluid filling needle injections for gradual stretching of the above skin), the skin that covers them should be meticulously cared of. The skin ought to be filled gradually under control to prevent damage to the surrounding tissues and compli-



Figure 7. Tying conjoined twins into the erect position in special “tailored” chairs

cations that hinder separation surgery. Possible reddening or inflammations should be checked for daily. Moisturising ointments are beneficial, which prevent excessive dryness of the tense, thinned skin. Children should be positioned in such a way as to avoid or at least reduce the compression of expanders. Additionally, gel mattresses, pillows or other protecting materials are recommended for this purpose. The patients' positioning has to be changed from the recumbent to the sitting position. In our centre, special “tailored” chairs were designed that enable for a maintained vertical position without compressing the expanders. Such measures are important not only for nursing procedures but they also help in proper personal development, contact with the surrounding environment and in individual trait development in the children [1, 3] (Fig. 7).

After separation surgery, it is recommended to continue invasive vital function monitoring in the twins. To restore and maintain homeostasis, children are clinically assessed for several hours after separation, whereby their biochemical and haematological parameters are controlled according to the accepted standards. After extensive separation surgery lasting for several hours, a postoperative pain management protocol that is previously prepared should be followed. In cases when simultaneous integument closure is not possible and artificial materials are used, continuous analgesedation



Figure 8. Dressing changes in the separated conjoined twins

should be administered to enable for supportive ventilation due to the unstable skeletal chest scaffolds [3, 16]. After separation of conjoined twins, the risk of multiple organ failure is higher, especially during the first postoperative days, which is attributable to the surgery extent and its long duration. Developing multiple organ failure symptoms should be diagnosed early and suitably treated.

Homeostasis maintenance is of particular importance in separated conjoined twins, especially in children with incomplete integument closures. Frequent surgical interventions for changing the artificial materials that cover the defect as well as dressing changes lead to overcooling of the separated children. Warming mattresses and external radiators allow for constant body temperature maintenance. Moreover, it should be remembered that the large operative wound surface, together with the many invasively placed vascular lines, drains and catheters favour infection development. Conjoined twins with developmental intestinal and urogenital defects are prone to infections that can lead to sepsis [17]. Unfortunately, the majority of early complications and deaths after pre-separation and separation surgeries are caused by sepsis [9, 10, 18].



Figure 9. *Dicephalus dipus dibrachius* twins

During the first postoperative days, the gastrointestinal tracts of these children are not efficient enough to ensure for proper amounts of calories; therefore, parenteral nutrition is necessary [3, 16].

After separation surgery, children are subjected to frequent dressing changes and undergo consultations and interventions by the various teams involved in the separation and postoperative management. The interventions by the individual teams should be synchronised to allow for the children to rest. Once deep sedation is withdrawn and the digestive systems are mobilised, the 24-hour feeding, wakefulness and rest rhythm should be restored. Endotracheal tubes and vascular accesses should also be controlled, whereby respiratory and general rehabilitation management is required [3] (Fig. 8).

In recent Cracow centre cases, conjoined twins who did not undergo separation procedures were treated in the ICU using all of the available methods. The twins with additional congenital defects died shortly after birth despite the intensive therapy applied. The remaining twins were treated for several months and all of the encountered problems were identical or similar to those discussed above (Figs 9, 10).

The proper psychomotor development of infants and small children is conditioned by the appropriate inflow of external stimuli. Conjoined twins cannot be carried in the arms as healthy infants can. Thus, their proper develop-



Figure 10. Thoraco-omphalopagus twins



Figure 11. Conjoined twins — one organism, two different personalities

ment depends on limited stimuli from the nearest surroundings. The ICU personnel are obliged to provide such stimuli. The activities vital for the development of a child, such as changing positions and sitting or standing, are not possible or are markedly limited in conjoined twins. Early and symmetrical rehabilitation is also important, in which suitable exercises should help develop and strengthen their muscles. Some conjoined twin types (e.g., *thoracopagus* twins) require special devices that support them in the sitting position. Specifically, supporting pillows can be used for this purpose. Special supportive devices that enable for standing are necessary for *ischiopagus* twins. Upper limb motor coordination in conjoined twins is usually normal as the majority of them have normally developed upper limbs. In most cases, this is the only possible method of physical contact with the surroundings. Generally, the development of speech and emotional reactions of conjoined twins is normal, if verbal and visual communication is ensured.

Although their genetic code is identical, conjoined twins have different personalities (Fig. 11).

Separation surgeries that are too long or postponed can inhibit individual personality development in these children [18, 19, 20]. After separation, the children should stay near to each other [3].

Another important issue in conjoined twin therapy is associated with psychological-ethical aspects. For many weeks, months, and sometimes even years, the team that is attending the Siamese twins establishes special and close connections with the children. The exact understanding of the “uniqueness” of the conjoined twins triggers much more involvement by the medical personnel than in other cases involving children. This involvement is reciprocal. Biological parents do not spend time with them every day, sometimes for reasons that are not dependent upon them. In such

cases, the attending team is also particularly affected by each treatment failure [4].

References:

1. Grochowski J, Bysiek A, Górska M *et al.*: Problemy chirurgiczne i anestezyjologiczne w leczeniu trzech par zrosłaków. Pamiętnik 54 Jubileuszowego Zjazdu Towarzystwa Chirurgów Polskich, Kraków 1989: 388–392.
2. McDonald KM, McIntyre P, Hegney D: Separation of conjoined twins: experiences of perioperative nurses and their recommendations. International Council of Nurses, Int Nurs Rev 2005; 52: 52–59.
3. Jakobowski DS, Hagelgans NA, Leson JK: Assessment and management of conjoined twins. J Perinat Neonatal Nurs 1989; 3: 66–82.
4. Pierce SF: Neonatal intensive care. Decision making in the face of prognostic uncertainty. Case: Conjoined twins. Nurs Clin North Am 1998; 33: 287–297.
5. Wirt SW, Algren CL, Wallace VR, Glass N: Separation of conjoined twins. AORN J 1995; 62: 527–545.
6. Hobaike ABS, Pires KCC, Fernandez VB SA: Anesthesia for emergency separation of omphalopagus conjoined twins. Rev Bras Anestesiologia 2010; 60: 311–314.
7. Diaz JH, Furman EB: Perioperative management of conjoined twins. Anesthesiology 1987; 67: 965–973.
8. Ibinaiye PO, Mshelbwala PM, Abdulgafar N, Lawal AK: Dicephalus dipus tetrabrachius conjoined twins of Zaria: Case report and literature review. Niger J Clin Pract 2013; 16: 395–397.
9. Thomas JM: Anaesthesia for conjoined twins. Childs Nerv Syst 2004; 20: 538–546.
10. Singh M, Jacob R, Naik V, Baines D: Separation of thoraco-omphalopagus twins in a rural secondary hospital: perioperative management. Indian J Anaesth 2012; 56: 442–447.
11. Fisher K, Throne M, Choudhry D *et al.*: Pediatric tracheostomy in conjoined twins: Multidisciplinary approach. Int J Pediatr Otorhinolaryngol 2009; 73: 487–491.
12. Geoges LS, Smith KW, Wong KC: Anesthetic challenges in separation of craniopagus twins. Anesth Analg 1987; 66: 783–787.
13. Harrison VL, Keneally JP, Gold PD, Malcolm PS, Overton JH: Anaesthesia for separation of conjoined twins in the neonatal period. Anaesth Intensive Care 1985; 13: 82–85.
14. Shank E, Manohar N, Schmidt U: Anesthetic management for thoracopagus twins with complex cyanotic heart disease in the magnetic resonance imaging suite. Anesth Analg 2005; 100: 361–364.
15. Skoczylas-Stoba B: Utrzymywanie drożności dróg oddechowych. In: Szreter T (ed.): Intensywna terapia dzieci. Wydawnictwo Lekarskie PZWL, Warszawa 2002: 69–78.
16. James PD, Lerman J, McLeod ME, Relton JES, Creighton RE: Anaesthetic considerations for separation of omphalo-ischiopagus tripus twins. Can Anaesth Soc J 1985; 32: 402–411.

17. *Wilcom DT, Quinn FM, Spitz L, Kiely EM, Ransley PG*: Urological problems in conjoined twins. *Br J Urol* 1998; 81: 905–910.
18. *Chalam KS*: Anaesthetic management of conjoined twins separation surgery. *Indian J Anaesth* 2009; 53: 294–301.
19. *Khan ZH, Tabatabai SA, Saberi H*: Anesthetic and surgical experience in a case of total vertical craniopagus. *Surg Neurol* 1999; 52: 62–67.
20. *Aoun Tannuri AC, Pereira Batatinha JA, Prieto Velhote MC, Tannuri U*: Conjoined twins — twenty years experience at a reference center in Brazil. *Clinics* 2013; 68: 371–377.

Corresponding author:

Krzysztof Kobylarz, MD, PhD
Department of Anaesthesiology and Intensive Therapy
University Children Hospital
Collegium Medicum, Jagiellonian University
ul. Wielicka 265, 30–663 Kraków, Poland
e-mail: kkobylarz@usdk.pl

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