Specificities of Nursing Care of Conjoined Twins

Abstract

Conjoined ("Siamese") twins are one of the rarest congenital anomalies with an incidence of about 1.47 per 100,000 births in the Western world. This rare natural phenomenon is present in only 1% of monozygotic twins, i.e., 0.05% of live births. Most of them are stillborn (40-60%) or die early in life. The majority of live births are female (75%), which is why the female karyotype is considered to be beneficial in terms of survival. Conjoined twins are created when the zygote begins to separate after 13 days post-fertilization, at a time when the so-called embryonic disc has already been formed. Instead of producing two separate embryos, this late separation causes incomplete separation of the embryos. The name comes from conjoined twin brothers Chang and Eng Bunker, who were born in Siam. Surgical separation of conjoined twins is usually a very demanding and complex procedure, and often not even possible. Survival after separation depends on the type of connection.

This paper presents the case of conjoined twins who were born at 33 weeks. At 11 weeks of pregnancy, the doctors suspected that the twins were joined ventrally (omphalopagus - twins joined at the abdomen). The diagnosis was confirmed by fetal MRI at 21 weeks. Separation was indicated at the age of 45 days and a combined body weight of 4,700 g. Long postoperative recovery and treatment lasted three months and was complicated by infections and problems with the healing of the surgical wound of one of the twins. Treatment and recovery required a multidisciplinary approach and a well-educated team of doctors and nurses.
Introduction

Conjoined twins are twins who are physically fused in the womb and consequently at birth. The first mention of conjoined twins dates back to the Neolithic period (1). The most common etiology of conjoined twins is partial fission or secondary fusion. Both of these processes can be used to explain the embryological and anatomical findings observed in conjoined twins (2). Conjoined twins are the result of axis splitting or duplication that occurs after 13 days post-fertilization. They are described according to the site of fusion and are a rare atypical presentation of symmetric monozygotic twin pregnancies. They are of the same sex and karyotype. They are more common in women with a ratio of men to women of 1:3. They are the result of incomplete embryo splitting into two separate twins after 12 days post-fertilisation. Conjoined twins are more often female. They are classified according to the point of fusion: ventral-rostral, caudal, lateral and dorsal. Among conjoined twins, Thoracophagus is the most common type (3,4). Traditional healthcare strategies may require modification to meet the unique care needs of conjoined twins. Areas of focus include ensuring privacy, designing appropriate housing units to meet space and equipment needs, staffing considerations, and adaptations to typical neonatal intensive care interventions. Caring for conjoined twins is a complex process and the role of the nurse has an important impact on the overall outcome. Even basic and routine nursing tasks must be adapted to meet the needs of these unique patients (5).

Conjoined twins

Conjoined twins are a rare embryological developmental disorder of unclear etiology. The condition itself implies malformations and is associated with secondary changes associated with abnormally connected organs and the superimposed effects of abnormal hemodynamics. The proposed defect mechanisms cannot explain the changes in the normal developmental process, whereby a pair of monozygotic twins do not separate from each other and continue their normal embryological development (6). The rarity and complexity of this condition present a challenge for prenatal planning and postnatal care. Medical, imaging, and surgical advances have led to earlier, more accurate depictions of anatomy, and as a result, have allowed more time for the team of professionals to prepare for their complex care (7).

Historical background

Since ancient times, conjoined twins have captured the attention and fascination of people around the world. Over the centuries they have been worshipped as gods, feared as monsters, and paraded before curious crowds for entertainment. They were integrated into the myths and legends of many ancient civilizations, especially Janus, the ancient Roman god of beginnings and ends. For centuries, people considered conjoined twins, as well as children with birth defects, to be a bad omen or a sign of God’s wrath (8). Probably one of the first documented cases was
a pair of conjoined twins born in Isle-Brewers, England, joined at the back from the middle of the chest to near the lumbar region part. Anecdotal reports of live conjoined twins in European medical history date back more than 1,000 years, but the first well-known case was not documented until 1811 when 2 boys - Chang and Eng - were born in Bangkok, Thailand, attached at the sternum and they were called “Siamese twins” (Figure 1). As they travelled the world with Barnum’s Circus, they consulted with a multitude of doctors. Everyone, including Rudolf Virchow, concluded that separation would be fatal for both of them (9).

Epidemiology
The exact incidence of conjoined twins has not been determined, and the estimated frequency varies in the literature. All conjoined twins are monoamniotic, monochorionic and monozygotic twins. Conjoined twins occur in 1.6% of all human pregnancies, of which 1.2% are fraternal and 0.4% are monozygotic. Monochorionic-monoamniotic twins account for less than 1% of identical twins, and conjoined twins are even rarer, occurring in approximately 1 in 50,000-100,000 and 1:600 live births. Conjoined twins are three times more common in female fetuses than in male fetuses. It is believed that 1 in 40 monozygotic twins fail to separate, producing conjoined twins. There was a reported incidence of 1:14,000 live births in India and Africa and 1:250,000 live births in Europe and the USA, suggesting an increased incidence in the black population (10). More recent studies on the epidemiology of conjoined twins are relatively rare, but the prevalence does not appear to be significantly different (1.02-1.34 per 100,000 births) in Western populations. However, an increased prevalence of 3.27:100,000 births and 2.85:100,000 births was reported in two studies on the Chinese population from the same Surveillance Program at different times (6).

Embryology
The anatomical structure and physiological functioning of the female reproductive system are predisposed for the conception of one embryo which, after implantation, develops into a fetus and is born naturally as a normal newborn at full term. Sometimes, however, various disorders can disrupt this sequence of processes. A special example of such a disorder is multiple pregnancies, which are often and incorrectly called twin pregnancies because there can be more than two fetuses. Ovulation disorders can lead to the release of several eggs that, when joined with sperm, form independent zygotes and then polyzygotic fetuses. Embryonic regulation is a specific characteristic of early embryo development. It can lead to the development of genetically identical (monozygotic) twins. This phenomenon is not treated as a pathology, especially since further embryonic development, birth and postnatal development are often normal (11). It is now generally accepted that a human embryo can divide and form into monozygotic or identical twins at one of three stages of development. After fission or cleavage, two genetically identical embryos would be expected and after zona lysis, each would implant and develop as two distinct, although genetically identical, embryos (12).

Currently, the generally accepted and prevailing theory that could explain the origin and development of fused fetuses is the fission theory. It is assumed that during gastrulation when the embryo still has the shape of an elliptical disc, the disc spontaneously cleaves incompletely along the cephalocaudal axis. The process of embryonic regulation creates conjoined twins. They are characterized by homologous connection, which means that both individuals share the same organs, e.g., head-to-head or back-to-back. Fragmentation of the embryonic disc and separation of tissue elements can be caused by environmental factors, cell adhesion disorders or increased genetically conditioned apoptosis, i.e., cell death. An alternative theory that explains the origin of conjoined twins is the spherical theory. It is assumed that the embryonic discs, which are formed after the complete cleavage of the embryo in the early stage of development, may have a common yolk sac. The cells that form it can “swim” or “float” on its surface and rejoin, usually endodermally, thus creating ventrally conjoined twins. On the other hand, dorsally conjoined twins are the result of ectodermal fusion due to the approximation of embryonic discs that share a common amniotic cavity at an early stage (11).

Classification
The most common case of conjoined twins is two conjoined children of a similar stage of development. In general, conjoined twins are divided into ventrally fused conjoined twins (approximately 69.3% of cases) and dorsally fused conjoined twins (5.4%).
The remaining types include connections of a non-specific nature (21.4%) and the so-called parasitic twins (3.9%), i.e., those characterized by significant morphological disproportions. The nomenclature associated with the pathologies observed in the case of joined fetuses is based on the morphology of the connection. Ventrally conjoined twins can be divided into cephalically conjoined, which include thoracopagus (42%), cephalopagus (5.5%) and omphalopagus (5.5%). An example of caudal fusion is ischiopagus (1.8%), and lateral fusion parapagus (14.5%) (Figure 2) (13).

Ventrally fused conjoined twins initially share a common yolk sac that participates in the development of the intestines, liver and pancreas. They usually share a common peritoneal cavity and umbilical cord. Their hearts can be shared or separated depending on the level of fusion. The thoracopagus type is formed by fusion at the cardiac primordium. The connection includes the chest and the upper part of the abdominal cavity. In this case, numerous common organs prevent separation. The cephalophagous type develops by initial fusion on the oropharyngeal membrane at the anterior part of the embryonic disc. After that, it stretches from the top of the head to the navel. In this case, the trunks of both fetuses share the head and chest. The prognosis for their survival and development is very unfavourable (1.2). The omphalopagus type develops by the fusion of the primordial diaphragm. The connection includes the chest and the upper part of the abdominal cavity. The ischiopagus type is associated with a common cloacal membrane and the development of the hindgut and rectum. It also includes the musculoskeletal system of the back of the body. Such fusion can also be found in the lower part of the abdominal cavity and the area of the urogenital tract. Fusion usually does not involve critical organs and newborns can be effectively separated. The parapagus type develops by fusion of the anal primordium, often retaining two notochords, and eventually the lower spinal segments. The connection involves the pelvis and torso to varying degrees; there are one or two heads and two faces (13). Dorsally conjoined twins are divided into the following types: craniopagus (3.4%), rahipagus (1.0%) and pygopagus (1%). These twins share a part of the axial nervous system that originates from the neural tube (Figure 3) (7).

The spine and its muscles are common, but there are two abdominal cavities. Umbilical cords are usually separate. The craniopagus type develops with an
initial fusion in the region of the anterior neural foramen that ultimately includes the cranial vault. The rachipagus type develops from the fusion of the central part of the neural tube that includes the spine and its muscles. The pygopagus anomaly is formed by the fusion of the caudal segments of the neural tube in the area of the posterior neural opening and ultimately involves the sacrum and coccyx with the pelvic muscles. It is important to emphasize that all types of conjoined twins have a common etiology (7).

Prenatal diagnosis

Developments in imaging and radiology have enabled physicians to diagnose and assess the extent of fusion at an early developmental stage in most cases of conjoined twins. It is now possible for the medical team to more accurately predict the outcome and adequately counsel families to help them decide whether to continue with the pregnancy or opt for early termination. Prenatal assessment is currently primarily performed using ultrasound, with the addition of colour Doppler imaging and a three-dimensional ultrasound, which help in establishing an accurate diagnosis (8). Currently, prenatal ultrasound is commonly used to diagnose conjoined twin pregnancies and can be performed as early as 12 weeks of pregnancy. However, a detailed scan at 20 weeks gestation gives a reasonable estimate of the extent of the fusion and any other problems. The non-invasive nature and safety of this procedure make it the modality of choice for early in-utero diagnosis. This technique uses the amniotic fluid’s ability to act as a buffer during scanning. When appropriate, transvaginal ultrasound is also recommended. According to recent studies, prenatal fetal MRI can add information to help define the exact anomalies and connect anatomies between fetuses, as a result of its higher resolution, lack of ionizing radiation and its non-invasive nature. They are widely used to delineate anatomical details, especially the cardiovascular system. A detailed examination of the heart is particularly important because the prevalence rate of congenital heart abnormalities in conjoined twins is high (14). Correct diagnosis is also an important aspect in planning the obstetric care of conjoined twins, as are the decisions about the method and timing of delivery. Early diagnosis of conjoined twins will then allow for better management including preventing premature birth and allowing the fetuses to continue their normal development. The method of choice for delivering conjoined twins is a Cesarean section. Needless to say, the more one knows about the twins before the operation, the better the outcome of the separation procedure will be (15).

Treatment

If the diagnosis is made in the first trimester, the twins should be delivered by a Cesarean section at or near the hospital where the separation will take place. However, in many cases, the condition is not discovered until labour is obstructed (16). Conjoined twins fall into one of three groups - namely newborns who do not survive birth (and those who die shortly after birth), infants who survive till elective surgery and infants who require emergency surgery. Elective separation is possible in omphalopagus, pygopagus and some craniopagus, and thoracopagus twins. Separation is not possible with cephalopagus, parapagus and rahiopagus twins. Urgent separation may be necessary when there is damage to the connecting bridge (omphalopagus), where one twin threatens the life of the other (complex congenital heart disease, cardiomyopathy, sepsis), when the condition of both twins deteriorates due to hemodynamic and respiratory disorders (typically thoracopagus) or where the condition of one of the twins is incompatible with life (anencephalic, stillborn), and the other has good prospects for survival (17).

As a result of the multiple organ abnormalities of conjoined twins, it is necessary to perform a thorough examination of each organ system. The most critical organ that determines the prognosis of surgical separation is the heart. Parts of the gastrointestinal (GI) system are usually shared in most cases of conjoined twins, except in craniopagus twins. An adequate examination of the digestive tract can be done by applying dyes at a separate time intervals to understand which structures are being divided. Ultrasound, 3D-MRA and radioisotopes can be used for analysis in the presence of a common pancreas and biliary system, which are often found in thoracopagus and omphalopagus twins. Cystoscopy has become a popular approach to examining the genitourinary system in cases involving the common bladder, rectum, and/or vagina. It is also important to assess the kidneys, including their number, location, and degree of fusion. Ultrasound and MRI are commonly used for such purposes. In male twins, joint use of the genitals, scrotum and testicles should be evaluated (18).
During the separation procedure, great attention is paid to anesthesia, breathing and hemostasis. Certain experts are assigned to each twin separately, along with a joint team coordinator. Due to shared circulation, poorer cardiac function, and unpredictability of reactions to drugs, administration of anesthesia during the separation procedure can be difficult. Endotracheal intubation is strongly recommended to maintain adequate respiratory gas exchange. However, it can be difficult to intubate thoracopagus twins (10). Cross-circulation between conjoined twins has significant implications for anesthetic pharmacology and resuscitation. Where cross-circulation is imbalanced, significant end-organ (cardiac, renal, and respiratory) failure can occur. It should also be recognized that the degree of cross-circulation is dynamic and depends on the relative systemic vascular resistance of both twins. Although dosing of anesthetics, antibiotics, and analgesics is often based on weight (using half the combined weight as a standard), effects can be erratic. Induction of anesthesia should be simultaneous since inhalation of volatile substances can cause complete, no or some effect on the other twin.

Heparin dosing before cardiopulmonary bypass may be compromised by unequal flow distribution between the twins. Careful angiographic imaging of the cross-circulation is necessary to assess the fraction of cardiac output shared between twins because the survival of one twin may depend on the circulation of the other (16). Due to the possibility of large blood loss, it is very important to maintain adequate tissue perfusion. Conjoined twins usually have critical energy needs due to significant cardiopulmonary and metabolic abnormalities. Therefore, dextrose and isotonic electrolyte solutions can be administered intravenously. When necessary, palliative procedures including amputations, draining colostomies and ileal conduits are performed. Complications such as arrhythmia, cardiac arrest and vascular thrombosis may occur during surgical procedures. There is also an increased risk of anesthesia complications, hypothermia, and ventilatory failure due to the need for prolonged anesthesia. Greater radiation exposure and a greater need for contrast administration also require careful consideration (19).

After the surgical separation, the twins need intensive treatment and care. They should be transported to intensive care units and constantly monitored for signs of bleeding, hypotension, hypothermia, hypocalcemia, electrolyte loss, hypoxia, and acid-base imbalance. Cardiovascular, pulmonary and renal functional indicators should be continuously recorded to prevent any complications. Mechanical ventilation may be necessary for twins with severe respiratory distress. A review of the literature indicates that cardiovascular and respiratory failure are still the two major risk factors for twin death after separation. The stress of long-term treatment and surgery puts the twins at risk for infections and delayed wound healing. Therefore, central venous lines should be maintained for rapid access to provide intravenous boluses of fluids, blood, parenteral nutrition, or anesthetics. Additional surgical procedures may be required to repair any other visceral abnormalities (8).

**Ethical aspects**

Health workers often face ethical dilemmas in their practice. However, conjoined twins bring their own unique set of ethical challenges. Already at the initial diagnosis, the decision to continue or terminate the pregnancy is often difficult. However, the real challenge begins at birth, when the decision is made for or against separation. The main goal of the separation procedure is not only to save lives but also to improve the quality of life of the twins. Except emergencies that require separation, this notion assumes that twins do not have a fulfilling life if they are not separated (10). Cases where there is significant organ sharing, yet two different functioning brains, are philosophically and ethically challenging. This is because they raise questions about a person’s identity, whether they are identical to something psychologically or biologically, they force a person to decide whether what matters from an ethical point of view is the biological life of organisms or the existence of consciousness or mind. They raise questions about when, if at all, it is morally acceptable to sacrifice one person to save another. They force you to think about the conditions of organ ownership and the justification of organ removal for transplantation that causes the death of the donor. They raise questions about who should make decisions about life-threatening treatments when patients themselves cannot decide (20). These cases raise serious moral issues about whether surgical separation should be attempted when extensive organ overlap makes the conjoined twins’ life prospects poor, but their surgical separation is medically difficult and risks worsening the life prospects of at least one of the twins (14). If the necessary operation is easily performed and without
risk, there is a strong moral reason to perform it, since it is likely to improve the life prospects of the twins even more, since conjoining in all its forms brings some disadvantages, such as reduced privacy and exposure to public opinion. curiosity. Such cases can conditionally be called win-win cases because the life prospects of both twins are improved by separation (21). This case report was conducted with the permission of the parents and in accordance with the guidelines for the safety of persons participating in such research, including the Declaration of Helsinki.

Specificities of nursing care of conjoined twins

The specificities of nursing care of conjoined twins will be shown in the example of the girls born in the 33 weeks of gestation who were admitted to the Department of Neonatology and Neonatal Intensive Care of the Zagreb Clinical Hospital Center 65 minutes after birth. Treatment and care for conjoined twins involve patient-centred care, a multidisciplinary team consisting of neonatologists, intensivists, surgeons, cardiologists, radiologists, pulmonologists, gastroenterologists, and a nurse is an indispensable member of this team. Likewise, caring for conjoined twins requires continuity of care, consistent communication, and the development of trust and comfort between staff and family.

Prenatal diagnostics

The girls V. and K. T. were born from the third, spontaneously conceived and regularly controlled pregnancy in which a twin pregnancy was monitored from the 11 weeks gestation when it was suspected that they were twins joined in the abdominal area. At 21 weeks gestation, a fetal MRI was performed, when two fetuses were confirmed to be joined ventrally by a common liver (fused in the abdominal area with the largest craniocaudal diameter of 4 cm - omphalopagus type). The thorax and pelvis of the fetus were distinctly separated. Every fetus had properly developed upper and lower limbs. Separate stomachs were observed, but this method cannot confirm or exclude the existence of a common biliary system (gallbladders were not observed with certainty) or other parts of the alimentary canal. Dilation of the intrahepatic bile ducts was not observed. The diaphragm was not visible as an anatomical structure, but its development was certain given the clear separation of the anatomical structures of the abdomen from those of the thorax, without signs of diaphragmatic hernias. Nevertheless, a common diaphragm in the ventral part of both fetuses could not be ruled out. A separate urinary bladder was visible in both fetuses, as well as separate hearts with a regular display of collapsed lung parenchyma. As far as was available for analysis, there was one placenta, and one umbilical cord that was traced to the caudal part of the ventral junction. With this presentation, it was not possible to analyze the details of the blood flow of the fetuses, that is, to confirm or exclude the involvement of large blood vessels, especially veins, in the area of the omphalopagus. The presentation of cord parenchyma of both fetuses was regular.

Fetal echocardiography in the 29 and 33 weeks gestation showed regular morphology of the hearts of both fetuses. On 10 January 2019 (GD 32+3 weeks), the mother was hospitalized at the Department of Pregnancy Pathology of the Sveti Duh Clinical Hospital due to planned preparations for the termination of pregnancy, and on 16 January 2019 at 9:41 a.m., 2 female premature babies were born by Cesarean section, fused in the area of the lower part of the thorax and abdomen, with a combined birth weight of 3800 grams. The first twin was 43 cm long, with a head circumference of 29 cm, Apgar 7/8, cyanotic, not breathing on her own, with a heart rate <100/min, intubated, ventilated manually with a self-expanding balloon, and a peripheral venous line was installed. The second twin was 41 cm long, with a head circumference of 30 cm, Apgar 6/7/8, cyanotic, not breathing on her own, with a heart rate <100/min, intubated and ventilated manually with a self-expanding balloon, and a peripheral venous line was placed and adrenaline administered due to bradycardia. For further treatment and care, the conjoined twins were transported in a transport incubator to the Clinical Hospital Center Zagreb, Department of Neonatology and Neonatal Intensive Care Medicine.

Admission of conjoined twins

Upon arrival at the department, the first twin, V.T., was orally intubated, manually ventilated, eupnoeic, did not use auxiliary respiratory musculature, had little spontaneous motor activity, blood oxygen saturation (SpO2) was 90%, heart rate 140/min, blood pressure 45/24 (33) mmHg, and body temperature measured cutaneously was 35.1°C. The second twin, K.T., was orally intubated, manually ventilated, cy-
Preoperative care

The twin girls were placed in an incubator and connected to mechanical ventilation. The most commonly used vascular approaches in neonatal care are the peripherally inserted central catheter (PICC) and the umbilical catheter (22). Due to the obvious impossibility of placing an umbilical catheter, girls V. and K. were inserted with PICC catheters.

Trophic feeding was started on the fourth day using breastmilk, with a gradual increase in the volume of the meals, which both twins tolerated adequately. On the twelfth day after being born, due to the necrotizing enterocolitis in the first twin, the enteral intake of both twins was stopped and antimicrobial therapy was started, which was quickly followed by a good clinical response and normalization of inflammation parameters. Due to the intertwin transfusion syndrome (cross-mixing of circulation between twins via a portosystemic shunt in the common liver parenchyma, proven by MSCT angiography), progressive arterial hypertension and polyuria (diuresis 308 ml/day) were observed in the one twin, and oliguria (diuresis 42 ml/day) in the other twin.

Serum electrolytes, creatinine and urea, due to abundant mixing of blood, were repeatedly uniform and within reference values in both twins. However, creatinine clearance in twin K. on the 30th day of life was 54 ml/min/1.73 m², and in twin V. 32 ml/min/1.73 m². The twins were connected to a monitor and the heart rate, blood pressure, number of respirations and blood oxygen saturation of each twin were recorded in the nursing documentation, and the laboratory findings from the venous blood sample were regularly checked. Ultrasound of the abdomen and colour doppler of the splenoportal basin were performed, showing 4 kidneys without signs of hydronephrosis, 2 separate urinary bladders, 2 spleens, 1 liver with normal echogenicity without dilatation of the bile ducts, 2 portal veins and normal hepatic arteries and veins. The gallbladder was shown only in the second twin. As already mentioned, the girls were on mechanical ventilation, each on their ventilator parameters, and the nurse monitored the respiratory status every day and periodically, if necessary, performed aspiration of the endotracheal tube. Endotracheal tube aspiration is a routine and common procedure in the intensive care unit to remove secretions and maintain airway patency so that oxygenation and ventilation in an intubated child can...
be optimized. However, tube aspiration can cause hypoxia due to aspiration of oxygen from the lungs and alveolar collapse (24). Therefore, it is important to carry out the aspiration procedure in an adequate way. It is important to choose the appropriate size of the aspiration catheter, which should be one-third the size of the diameter of the inner lumen of the tube. Aspiration is performed by two nurses; one who performs aseptic aspiration and one that assists. The aspiration nurse advances the aspiration catheter to a predetermined length, ensuring that the catheter is only passed through the length of the tube. Applying negative pressure, the nurse gently rotates the catheter as they pull it out of the tube. The duration of negative pressure should not exceed 6 seconds to prevent hypoxemia. In order to prevent accidental extubation, the assisting nurse gently holds the child’s head in a still position and perfuses the child between two aspiration procedures with a manual balloon.

When the procedure is complete, the nurse reconnects the ventilator tubing. After that, the nurse aspirates the child’s nose and oral cavity to free them of secretions using a 6- or 8-gauge probe catheter for this procedure. It is important to observe the child’s physiological parameters after aspiration. If closed suction is used for aspiration, the procedure is performed by one nurse because the aspiration catheter is protected in a foil that ensures sterility and there is no need to separate the child from the respirator. The method and duration of the procedure are the same as for open aspiration (25).

On the fifth day after birth, following the application of surfactant, the girls were taken off the ventilator, and respiratory support was continued with non-invasive mechanical ventilation (Figure 5).

Non-invasive mechanical ventilation has been in use for pediatric patients for many years. Historically, continuous positive airway pressure and bilevel positive airway pressure modes have been used for respiratory diseases, including neonatal apnea, bronchiolitis, asthma, and pneumonia. The newest type of non-invasive respiratory support is the application of a high flow of air and oxygen to a nasal cannula (high flow nasal cannula - HFNC), which has gained popularity in the last few years and its use is justified in the literature. Studies have shown that this method of respiratory support can reduce the need for intubation and ventilation, reduce the length of intensive care days and increase the comfort of the newborn. The skin, especially of young infants on long-term therapy, may be compromised under the interface due to pressure on the skin. Prevention of skin damage can be achieved by using a skin protectant (26). Maintaining the skin integrity of conjoined twins can be challenging due to limited positioning and mobility. During the care of the twins V. and K., a foam mattress was used to protect the skin from pressure injuries, and the twins were successfully placed on their mother’s lap for bonding and social interaction. An important component of achieving the mothering role is bonding with infants through direct physical interaction, encouraging talk, touch, physical nurturing and holding as early and as often as possible.
With conjoined twins, enabling such interaction may be more feasible when they are younger and smaller in size, making the early days and weeks crucial for optimizing the mother’s confidence. Foam packs helped reduce the redness of bony prominences. Gel pads have also been useful during hospitalization in reducing pressure on bone prominences. Repositioning and turning are imperative to prevent pressure injuries and maintain comfort. The twins’ positions were changed every three hours. A key consideration in repositioning is maintaining twin identification. The twins needed to remain on their designated side of the bed to ensure proper identification. During hospitalization, the first twin V. T. urinated sparingly, had no bowel movement and developed necrotizing enterocolitis (NEC), which was treated conservatively. The second twin K.T. urinated profusely (dialyzes the first twin), and a native CT showed a larger heart compared to the first twin, which was a consequence of hemodynamics, and she developed hypertension. On February 20, 2019, the medical advisory board held a meeting with the parents with the aim of making a decision on separation, but the decision was not reached (Figure 6). Six days later, at the re-meeting of the council, an agreement was reached on the surgical procedure due to the threatening decompensation of the second twin’s heart, and the separation procedure was agreed upon for March 2, 2019.

The conjoined twins were transported to the operating room on March 2, 2019, at 7 in the morning (Figure 7).

Figure 6. The plan for separating the twins
Source: Author

Figure 7. Separation surgery
Source: Author
The first twin kept a common part of the intestine from the duodenum to the omphalomesenteric duct, and because of NEC, a subtotal colectomy was performed and an ileostomy was formed. The intestinal continuity was established after 2, and the final closure of the abdominal wall after 4 months. The second twin kept a part of the remaining intestine (the duodenum before the joint part was connected to the ileum, and the abdominal wall was closed using Vacuum-Assisted Closure (VAC). The separated twins were transported back to the Department around 10 p.m.

Nursing care after surgery

After being admitted from the operating room, the girls were each placed in their incubator and cared for by two nurses. Continuous monitoring and methods of intensive treatment were carried out, which included the use of antimicrobial and antifungal therapy, parenteral nutrition and nursing care. After separation, the other twin established adequate diuresis and gradual regression of arterial hypertension and hypertrophic cardiomyopathy occurred. During the course of stay after the operation, there was repeated appearance of fresh blood in the stool and abdominal distension, which normalized only after the use of therapy and a semi-elemental milk formula (Alfare).

During the care of the first twin, the nurse paid special attention to the surgical wound and ileostomy due to the development of possible complications (Figure 9). The appearance of the ileostomy was assessed by observing the colour of the stoma and the surrounding skin. A red or dark pink stoma indicates adequate blood supply, while pale pink indicates reduced hemoglobin or poor perfusion, and grey to black indicates ischemia and potential necrosis, and urgent surgical examination is required. It is also necessary to assess the protrusion of the ileostomy, which is normally 0.5 - 1 cm above the skin. A retracted stoma indicates complications and is located below the level of the skin, as well as a prolapse, which indicates a protrusion of the stoma more than 2-3 cm above the skin (27). An infant with a formed ileostomy requires frequent and regular assessment of fluid and electrolyte status to prevent complications. Sodium is crucial for growth and infants with persistent sodium deficit are at risk of reduced growth and cognitive dysfunction. It is necessary to check serum electrolytes, urea and creatinine at least once a day until it stabilizes, then once a week until reanastomosis. More frequent monitoring depends on the child’s weight gain, stoma losses, age, electrolyte stability and general condition. Sodium in the urine should be checked once a week. In infants with good renal tubular reabsorption, urinary sodium is the best measure of total body sodium and level of depletion. However, results can be misleading in premature infants and those with kidney disease. A low level of sodium in the urine (<20 mmol/L) is an indication to start taking nutritional supplements. However, most neonates with a stoma will need a supplement of 2-4 mmol/kg/day, which implies the addition of 3% sodium intravenously and modification of total parenteral nutrition (28). The introduction of enteral nutrition increases the production of content that flows out of the ileostomy and requires the protection of the skin with a stoma bag. For this purpose, bags with circular base plates are used, which can release the contents at the bottom. However, if greater convexity is required due to the indentation of the scars, the protective base plate can be cut to the desired size. The bags are not changed for at least 24 to 72 hours to preserve the integrity of the skin (29). Bags pointing downwards can become soiled with urine and can negatively affect the integrity of the skin and the

Figure 9. The first twin after arriving at the Department after surgery

Source: Author
adhesion of the plate. If the base plate and the bag are well attached and there are no signs of leakage, the bag can be opened from the bottom and the contents can be removed using a syringe and the bag can be rinsed to remove the unpleasant smell. During the change of the base plate and the stoma bag, the nurse will wipe the skin around the entrance of the stoma with sterile water and slightly soft gauze, assess the appearance of the stoma, dry the skin with dry gauze, apply a protective spray and reattach the new base plate and bag (27). Due to the transitory immaturity of the intestines, most premature infants receive parenteral nutrition in the first few weeks of life. However, providing sufficient protein and energy to maintain optimal growth in such infants remains a challenge. Due to the immaturity of the gut during the first postnatal weeks, gastrointestinal feeding is insufficient to cover such needs at first. Despite the faster daily increase in rations, full enteral nutrition was achieved only around day 22 of life in a cohort of preterm infants weighing less than 1000 g. Parenteral nutrition supplies preterm infants with nutrients, but it is far from approaching the complex composition of cord blood (e.g., in terms of lactate, growth factors) and should not necessarily mimic fetal nutrient supply, dramatic changes in metabolism that occur after birth. The use of parenteral nutrition can cause serious complications (e.g., sepsis, cholestasis, thrombosis) and often does not meet the needs of premature infants (30). Therefore, it is necessary to switch to enteral nutrition as soon as possible, which in the case of these twins was difficult due to separation surgery and incomplete intestines. During the administration of breast milk through a nasogastric tube, nurses continuously assessed the appearance and behaviour of both twins, measured the circumference of the abdomen, checked the gastric retention before each feeding and checked the appearance of the stool. The course of the first twin’s stay was marked by recurrent ileus, peritonitis and difficult
healing of the surgical wound. After the first operation, she initially tolerated food properly and expelled stool on the ileostomy, however, four operations were performed to treat the ileus. Peritonitis was manifested by elevated levels of inflammatory parameters, a distended and painful abdomen and food intolerance. The surgical wound was difficult to heal, which required the nurse to implement interventions to prevent infection and improve healing. The nurse, with instructions from the surgeon, cleaned the surgical wound every day by using an aseptic method, washing the wound with a sterile saline solution, covering the surface with an Aquacell dressing to improve wound healing, and placing sterile dry dressings on the outer part, which she fixed with adhesive tape. It is important to note that in such cases it was also necessary to assess the surrounding skin, which was exposed to the daily change of adhesive tapes, which increased the risk of additional damage to the integrity of the abdominal skin. The nurse assessed the appearance of the wound daily and recorded all changes in the nursing documentation in order to adequately monitor the progress of wound healing (Figure 10). Intestinal continuity was restored after 2 months, and the final closure of the abdominal wall after 4 months.

After 3 and a half months in the Neonatal Intensive Care Unit, the twins were transferred to the Neonatal Post-Intensive Care Unit, where enteral nutrition, parental education and health care continued (Figure 11).

**Education and parental support**

The birth of conjoined twins can lead to psychosocial stressors, religious dilemmas and relationships with the community. This can lead to serious disruptions in family dynamics, interactions between the medical
team and the family, and among the medical team itself. Nurses play an extremely important role in educating and supporting the parents since they spend 4 hours a day with them and through long-term hospitalization can assess all fears, doubts and needs of parents. The parents of the conjoined twins V. and K. T. have been involved in the care of their children from the beginning and, with the help of nurses, established an emotional and physical bond with them. They carried out the twins’ hygiene every day, especially after the separation and monitored the feeding of the children. The mother expressed and stored breast milk regularly so that it could be given to the twins (Figure 12). The parents were educated about the possible food intolerance and the recognition of symptoms, about the independent implementation of personal hygiene, and they were also offered the support of a priest.

During hospitalization, parents actively participated in all the interventions related to the provision of nursing care. Due to their poor financial situation, help was organized for them with clothes and equipment for girls, as well as housing equipment. After a little more than seven months, on August 29, 2019, the girls were discharged with recommendations for further monitoring of growth and development (Figure 13).

Through coordinated care, the multidisciplinary team proactively identified the potential challenges of caring for conjoined twins. Nursing interventions made it possible to improve the condition and support the parents, who left the hospital extremely satisfied and still come to the Department every time the twins are scheduled for their follow-up appointment.

**Conclusion**

Conjoined twins, also known as “Siamese twins”, are a unique type of monozygotic twins and constitute an extremely rare condition that is often incompatible with life. However, there are types of conjoined twins in which both twins cannot be separated and survive. Continuity of care for conjoined twins requires the collaboration of a large number of health professionals who must work together to plan interventions and respond to the unique challenges they face. Coordination of this care should be seamless and requires regular meetings of a multidisciplinary team that provides an effective approach to coordinate interventions and address all the unique challenges faced in the care of conjoined twins. Nurses caring for conjoined twins must demonstrate autonomy, ingenuity, and passion in promoting optimal outcomes for these rare and complex patients. The paper presents the case of conjoined twins, the girls V. and K. who were admitted to the Department of Neonatology and Intensive Care Medicine after being born in the Sveti Duh Hospital. After a multidisciplinary assessment and diagnosis, it was decided to carry out the separation procedure. The procedure lasted 15 hours and was without complications. Caring for conjoined twins requires the implementation of specific nursing care procedures that are an essential part of care and treatment. It is extremely important to know the characteristics and symptoms of this specific and extremely rare group of patients in order to be able to plan and implement specific interventions. Likewise, it is important to include parents in care as well.

**REFERENCES**

SPECIFIČNOSTI SESTRINSKE SKRBI U ZBRINJAVANJU SIJAMSKIH BLIZANACA

Sažetak

Sijamski blizanci jedna su od najrjeđih prirođenih anomalija s incidencijom u zapadnom svijetu od oko 1,47 na 100 000 poroda. Ovaj rijedak prirodni fenomen zastupljen je u svega 1 % monozigotnih blizanaca, tj. 0,05 % živorodnih blizanaca. Većina ih je mrtvorođena (40 do 60 %) ili umire rano u životu. Većina živorodnih ženskog je spola (75 %), zbog čega se smatra da ženski kariotip nosi korist u smislu preživljenja. Sijamski blizanci nastaju kada se zigota počinje razdvajati nakon 13. dana od oplodnje, u vremenu kada je već formiran tzv. embrionalni disk. Umjesto da nastanu dva odvojena embrija, ovo kasno razdvajanje uzrokuje nepotpuno odvajanje embrija. Naziv sijamski blizanci dolazi od sijamske braće Changa i Enga Bunkera, koji su rođeni u Sijamu. Kirurško razdvajanje sijamskih blizanaca najčešće je vrlo zahtjevano i složeno, a često nije moguće. Preživljenje nakon razdvajanja ovisi o vrsti i načinu spojenosti. U radu se prikazuju sijamske blizanke koje su rođene u 33. tjednu trudnoće. U 11. tjednu trudnoće postavljena je sumnja na ventralno spojivanje blizanki (omphalopagus – spajanje u području abdomena). Dijagnoza je potvrđena fetalnom magnetskom rezonancijom u 21. tjednu trudnoće. U dobi od 45 dana života i pri zajedničkoj tjelesnoj masi od 4700 g indicirano je njihovo razdvajanje. Dugi tijek poslijeoperacijskog oporavka i liječenja trajao je tri mjeseca i komplikirao se infekcijama i problemima cijeljenja operativne rane druge blizanke. Liječenje i oporavak zahtijevali su multidisciplinarni pristup i dobro educirani tim liječnika i medicinskih sestara.

Ključne riječi: sijamski blizanci, prenatalna dijagnoza, kirurško razdvajanje, multidisciplinarni tim, medicinska sestra