The Results Of Two Conjoined Twin Separations From January To June 2010 : Case Series

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Background: Conjoined twins are a rare resulting from late and incomplete division of monozygotic embryonic disk generally after 13th day of fertilization. Most cases of separation are extremely risky and life threatening.
Methods: We present two successful cases of conjoined twins separations, who were admitted to Cipto Mangunkusumo Hospital, both in early 2010. Lulu-Lala, the abdominophagus conjoined twins and Nayla-Nabila the Abdominothorakophagus conjoined twins.
Results: The successions of this conjoined twins separation depends on the techniques, precautions and the team that works together to treat the patients. One of the separation techniques is the utilization of tissue expander to loosen the skin and so can be close primarily by primary closure. The foremost precaution is the infection control, including preparation and sterilization.
Summary: Conjoined twins have a particular structural defect. Forty percent of them are stillborn and 35% survive only for one day. The mortality is increased by many causes and the most common cause is infection after surgery. All aspects in the team have to give priority to the sterility to avoid infection of those babies. The efforts to avoid the infections must have been done start from pre-surgery managements until the post-surgery.
Keywords: *conjoined twins, tissue expander, infection control, teamwork*

Latar Belakang: Kembar siam adalah kasus jarang yang terjadi, disebabkan keterlambatan dan tidak lengkapnya penyatuan lempeng embrio monozigot setelah hari ke-13 fertilisasi. Sebagian besar dari kasus pemisahan sangat beresiko dan mengancam nyawa.
Metodologi : Kami menyajikan 2 kasus kembar siam yang berhasil ditangani di RSUPN Cipto Mangunkusumo pada awal tahun 2010. Lulu-Lala, kembar siam dengan perlekatan abdominophagus, dan Nayla-Nabila, kembar siam dengan perlekatan Abdominothorakophagus.
Hasil: Keberhasilan dari pemisahan kembar siam tergantung pada teknik, persiapan dan tim yang bekerjasama untuk menangani pasien. Salah satu teknik pemisahan adalah menggunakan tissue expander pada kulit yang hilang sehingga dapat ditutup dengan penutupan primer. Hal yang terpenting untuk diperhatikan adalah pengontrolan infeksi, termasuk persiapan dan sterilisasi ruang rawat dan kamar operasi, memandikan bayi dengan antiseptik dan skrining tim.
Kesimpulan: Kembar siam mempunyai defek struktural yang khas. Empat puluh presen dari kasus kembar siam lahir mati dan 35% hanya dapat hidup satu hari.1 Mortalitas meningkat oleh banyak sebab dan penyebab tersering adalah infeksi setelah operasi. Seluruh bagian di dalam tim harus memberikan prioritas pada kesterilan untuk menghindari infeksi padi bayi.
Kata Kunci: *conjoined twins, tissue expander, infection control, teamwork*

Births of conjoined twins, whose skin and internal organs are fused together, are rare. Actual numbers for conjoined births vary from 1 in 20,000 to 1 in 100,000 pregnancies, and 40-60% are stillborn, with many others dying within the first few days after birth. In many cases, the surgery results in the death of one or both of the twins. Mortality rates for twins who undergo separation vary. Long term survival with or without separation depends on the anatomic site of attachment and the extent of shared organs. In many cases, the surgery results in the death of one or both of the twins.[1] The Queen Creek-area conjoined twins whose struggle for survival touched the hearts of ordinary people and movie stars alike, died Tuesday night in Seattle as surgeons tried to

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Tuesday night in Seattle as surgeons tried to make their shared heart work more efficiently by separations.[2] A one-week-old conjoined twin died following high-risk emergency surgery to separate her from her sister, a London hospital said on Wednesday Dec 3.[3] Sisters Ananda Oktavia Ramadani and Andini Oktavia Ramadina are born in East Java, Indonesia, on October 1. Conjoined at the chest and sharing a malformed heart, the sisters die during an operation to separate them at Public Hospital in Surabaya, East Java, on November 2.[4]

We present two successful cases of conjoined twins separations at Cipto Mangunkusumo Hospital, Jakarta. We used the same preparation for both twins. We prepared the doctors and nurses team consist of surgical specialists (pediatric and plastic surgeon), anesthesia specialist, pediatrician, a clinical microbiological and pathological specialist, child care nurses, ICU nurses, and operating theater nurses. Each team held their role in treating the patient. Pathological and microbiological teams plays their role in inspection of MRSA colonization in the twins parents and health care personnel by swabbing the place where most bacterial colonized (nose and armpits). The ICU and operating theater nurses, they sterilized the rooms, re-examine the function of positive pressure, air filter functions, checking the drugs, operating machine, equipment in PICU and operating theater. The patients were prepared for the surgery by fasting and bathed using betadine skin cleanser. Four hours before surgery, anesthesia team checked the room temperature, monitors, prepare the medications and calibrate anesthesia machine that will be used for both patients. Both babies were taken to the operating theater by the child care and medical team. Prophylaxis antibiotic then administered to both patients. After the induction by the anesthesia team, monitor the patients, sterilize the baby and the personnel by washing hands and wearing sterile gown, both patients are ready for surgery. The plastic surgeon team holds its role in defect closure. Primary closure is an option in this operation, for such purpose, in certain operation we use tissue expander to stretch the skin in order to close the defect.

CASE I: The Abdominophagus Conjoined Twins (Lulu-Lala)

Figure 1. Lulu and Lala, the abdominophagus conjoined twin.

The first case is a 7 months old conjoined twins Lulu-Lala (Figure 1), was referred from Soedarso Hospital, Pontianak. The mature born babies were both having female external genitalia and they had two separate heads, four upper and four lower extremities. They were attached at the anterior abdominal wall. The babies undergone a radiograph examination 9 days after admission and the radiograph showed abdominophagus with fused livers, hepatica arteries vascularization and portal vein fuses (Figure 2).

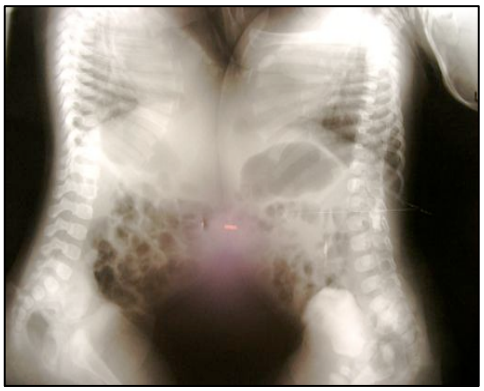

Figure 2. Radiograph of Lulu and Lala. It showed abdominophagus with fused livers, hepatica arteries vascularization and portal vein fuses.

Figure 3. Lulu After Separation

Figure 4. Lala After Separation

The operating team laid the patients to the operating table, washes it using povidone iodine then covered them with sterile fabric. The incision made by one of the pediatric team member, starting from the skin outer layer throughout the peritoneum. The livers were separated using harmonic scalpel. Ligations of the vascularizations and then the babies separated.

After doing the monitoring to both patients and in good condition, one of the babies is ready to be transferred to the operating table II. Baby Lulu is the one was transferred by the operating theater nurses. Babies who have been separated in each table, undergone pericardia fascia membrane closure by the pediatric surgeon. Now the plastic surgeon has their role. For Lulu (bigger baby), had been decided to close the defect by primary closure. The defect at the abdomen is too large to be close. We draw the bi-pedicle flap design and decided to make a vertical incision from inferior area of the right nipple to the RLQ abdomen. Then we performed STSG to close the incision defect (Fig.3). For baby Lala (smaller baby), primary closure can be performed without additional incision and decided to perform the Y inverted design (Fig.4). Operation completed.

CASE II: The Abdominothoracophagus Conjoined Twins (Nayla-Nabila).

The second case is Nayla-Nabila (Fig.5). The mother was G2P2A0 women and undergoing a routine follow up to Cipto Mangunkusumo Hospital at 20 weeks of gestation when conjoined twins were identified. Subsequently, ultrasonographic identified abdominophagus twins with four upper, four lower extremities, one organ for each baby and shared pericardium. The pregnancy continued until premature labor required hospitalization and ultimately cesarean delivery with premature rupture of gestational sac membrane as the indication.

Figure 5. Nayla and Nabila

Six months after delivery the babies undergone CT scan examination, some part of the Nabila's heart, mesenterium and their vascularizations are inside the Nayla's body (Figure 6). Some parts of Nayla's heart are inside the Nabila's body (Figure 7).

After some considerations and the calculation of the defect width, the plastic surgeon team decided to use tissue expander as a tool to loosen the skin that will be used to close the defect primarily (Figure 8).

The patients will be on their first operation, the insertions of tissue expander. After the patients was sedated, applied with the antiseptic and made an incision design, the pockets was made starts from both lateral and anterior sides of the chest. Operation completed after the implantation of the tissue expander and closing the incised skin. The initial

Figure 6. CT Scan With Contrast Nabilla.

Figure 7. Ct Scan With Contras Nayla

Figure 8. Nabilla-Nayla (Tissue Expander)

Figure 9. Tossue Expander Removed.

Figure 10. Nabilla After Separation

Figure 11. Nayla After Separation (Left)

Figure 12. Nayla After Separation (Right)

implantation for 200cc expander is 20cc and for the 30cc expander is 3cc. The next implantation for 200cc expander is 20cc and for the 30cc expander is 3 cc, 2 weeks after the initial. Then EMLA cream and wing needle are used for the remaining weeks. The total time required is 9 times implantation equal to 10 weeks.

After 1 month with tissue expander, Nayla got infected by pseudomonas and pneumonia. Tissue expander removed and patients were treated for 4 days (Fig.9). After the team meeting, the second insertion of tissue expander for Nayla had been decided. The patients undergone second operation. After 1 month of second expander insertion to Nayla, she got infected for the second times by pseudomonas with new problem, tissue expander exposed. At the end, the total volume expanders are 320cc for Nabila and 60cc for Nayla.

The infection treated, the team decided to do the separation surgery. After all the preparation, the operating team laid the patients to the operating table, wash it using povidone iodine then covered them with sterile fabric. The incision was made by the pediatric surgeon, at the abdomen area, and then continued to the diaphragm and thorax area, starting from the skin outer layer throughout the peritoneum (tissue expander maintained). After the incisions have done, exploring the ventricle and pericardium are the next step. Notified the attachment of both ventricles and pericardium, then they were separated bluntly. The livers were separated using harmonic

scalpel. The babies separated.

Defect closure at the fascia performed using appropriate pericardial membrane that has been made according to the design, and then sewn using prolene by the pediatric surgeon. Defect closure at the thorax area was closed by the thorax surgeon by notifying the rigidity of all ribs. Skin closure is the role of the plastic surgeon. All the tissue expanders removed. Nabila primary closure can be performed without additional incision and decided to perform the Y inverted technique (Figure 10). Nayla primary closure performed an additional contraincision at the left and right on lateral side of the abdominothoraco area (Figure 11 and Figure 12). The defects are too large to be closed. Then the closure defect of the incision (the right side) was then closed using STSG. Operation completed.

Observation of vital signs, ECG, urine production and saturation of pulse oxymetry is a must. Both patients transferred to the PICU. The team held a meeting to distribute watch over schedule for the babies. All team members have to be ready for any emergency call.

DISCUSSION

Conjoined twins represent a unique structural defect of monozygotic monoamnionic twins. The nonseparated parts of the otherwise normal twins remain fused throughout the remaining period of development. Forty percent of conjoined twins are stillborn, and an additional 35% survive only one day.[1] There are many causes increasing the mortality rate of the conjoined twin's separation. One of the primary causes is infection. Many twins died of infection after the surgery.[5, 6] The initial introduction of microbial pathogens occurs most often during the surgical procedure performed in the Operating Theatre (OT). Thus, to reduce the risk of SSI, a systematic but realistic approach must be applied with the awareness that this risk is influenced by characteristics of the patient, operation, personnel, and health care facility.[7] In handling those babies, we give priority to the sterility to avoid infection. Some efforts have been done starting from patients parents and medical personnel screening, equipment and operating theater sterilization, antiseptic babies bathing, and wearing the operating theater equipment for medical personnel. Declining the mortality Of the Two Conjoined Twins is also highly dependent with the personnel and the technique used in each operation.[8] In the management of those twins, a team was made by collecting a few personnel from their departments who are an expertise in their field. There's some provision to build a well organized team. They held meetings to make a planning series to organize in treating and operating those patients. We made the operating theater map completed with the machinery and personnel substitution to facilitate the movement in order to coordinate the surgery running smoothly (Figure 13). The personnel worked together very well as a team and using appropriate operation technique.

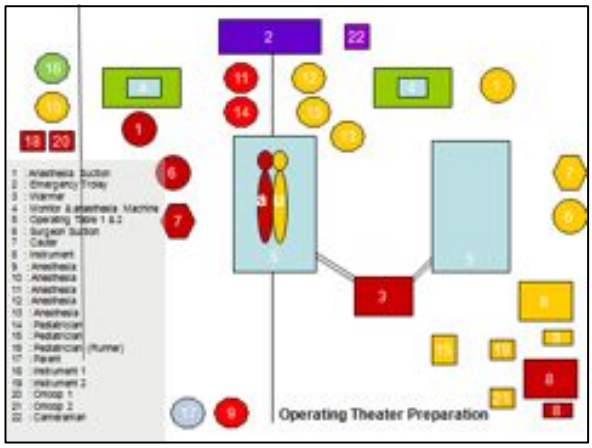

Figure 13. Operating Theater Preparation.

Figure 14. Tissue Expander

Plastic surgeons team plays their role in closing the defect. For the first case, Lulu's and Lala's were closed primarily, their skin are wide enough to close the defect. For the second case, Nayla's and Nabila's, the defect are too wide to be closed primarily. The plastic surgeons team tries to figure it out and found that using tissue expander is an appropriate choice. The tissue expanders are useful to stretch the skin then wide enough to close the defect. They used 2 two type of tissue expanders based on the volume, the 200cc and 30cc (Figure 14).

The tissue expanders filled gradually after being implanted. In such way, it will not make the skin stretched too vigorously that can cause pain. Infection happened to baby Nayla, tissue expander removed. After the infection treated, the team held meeting and decided to re-implant baby Nayla. Two months after the re-implatantion, she got infected for the second times by pseudomonas with new problem, tissue expander exposed. At the end, the volume of Nayla's tissue expander was smaller than Nabila's. On the day of separation surgery, it founded that the Nabila's skin can cover the defect by primary closure only and decided to perform the Y inverted technique. The Nayla's are too small and can't cover the whole defect. So the team decided to give a contra-incision on the left and right side of the abdominothoraco area. Then the closure defect of the incision (the right side) was then closed using STSG. Operation completed.

SUMMARY

Tissue expander in treating the separation of conjoined twins is one of the common chosen treatment. However, the infection control is the important part along the management of conjoined twins because it gives a big contribution for successfulness of the separation. Conjoined twins separations needs a full team who considers the importance of infection control.

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