Seminars in Pediatric Surgery 24 (2015) 231–236

Contents lists available at ScienceDirect

Seminars in Pediatric Surgery

journal homepage: www.elsevier.com/locate/sempedsurg

The separation procedure

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ARTICLE INFO

Keywords:
Separation of conjoined twins
Special cases
Results

ABSTRACT

The various stages of the separation are carefully planned but despite this, variations which will change the schedule of the procedure may exist. In general the operation commences on the opposite side from the main procedure and then the twins are turned for the remainder of the operation. Each type of conjoined twin is different but basically thoracopagus involves the hearts, omphalopagus involves the liver and small intestine and ischiopagus involves the large intestine and genito–urinary system.

Our results are presented together with interesting cases from which lessons have been learned.

Each type of union is associated with specific structural abnormalities—thoracopagus with cardiac union, omphalopagus with liver and intestinal fusion, ischiopagus the large intestine and genito–urinary system, pygopagus the perineum and terminal spinal cord and craniopagus the skull and meninges. Although structures may be joined this does not occur in a predictable manner. For instance, although liver fusion is frequently extensive, there may be one or two portal triads. In addition, although most of these infants have separate hepatic venous drainage, this is not invariably the case.

General considerations

The schedule of the operation is discussed in detail in the planning meetings. We have found that with almost all sets of twins, commencing the operation from the posterior aspect is preferable. This allows symmetrical division of the body wall and underlying structures as the initial incision directs the approach from the front. This entails turning the babies once they have been anaesthetised and intubated. As all the intravascular cannulas are in place, this can be a complicated manoeuvre.

We have never regretted undertaking this as division of the body wall is easier from outside than from within. In addition, with extensive union, it is difficult to maintain the correct plane of division and the body wall may not be allocated equally.

The incision posteriorly is carried down as far as seems advisable. The depth of the incision will vary—for instance, down to peritoneum when dealing with the abdomen and down to the sacrum with pygopagus twins. The object is to facilitate the direction of the approach from the front and to ease the final stages of the separation. This incision is closed with a running suture and the twins turned back for the main part of the operation.

Standard surgical techniques are used for separation. The tissues themselves are normal without any pathological process. The goal of surgery is to allocate organs fairly so each twin can survive independently.

The appearance after separation is often a surprise—the sheer size of the wound to be closed may be unexpected. In an attempt to provide more tissue for closure, we have used tissue expanders in the abdomen and found them to be troublesome and of little use. We are aware that others do not share this view but this is our experience. We are clear that expanders positioned over bone will give useful growth of skin and subcutaneous tissue and for this reason tissue expanders are very useful for craniopagus twins and for some pygopagus sets as well.

The method we have adopted entails the use of polypropylene mesh (Figure 1), attached to muscle, which is serially plicated to close the defect. Beneath the mesh we use a thin sheet of plastic (opened intestinal bag) to prevent adhesion of intestine and liver to the mesh during closure. The plastic is removed before the final closure. It is usual to achieve closure in less than 2 weeks. There seem few problems with the closure in later years. When planning the operation therefore, adequate sized mesh sheets must be available.

Thoraco–omphalopagus

On inspection, these may be identical. In some forms of omphalopagus twins, the bridge is very narrow and the variety of union quite obvious. However, by definition, the hearts are conjoined in thoracopagus but not in omphalopagus twins. If investigation suggests that the hearts may be separated, then the operation may proceed. The hearts share a common pericardial sac which may be divided, if sufficient tissue is available for each heart

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http://dx.doi.org/10.1053/j.sempedsurg.2015.06.007
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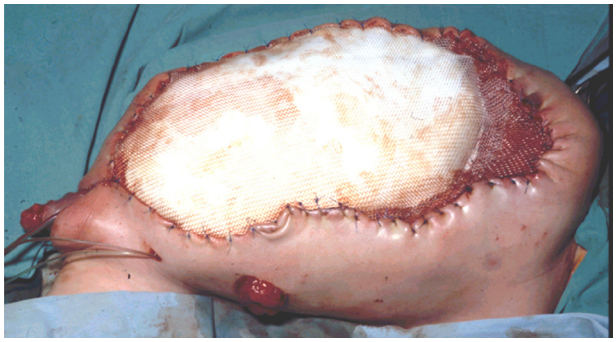

Fig. 1. Polypropylene mesh inserted in the abdominal wound for temporary closure.

(Figure 2). Alternatively, the defect in the pericardium will need to be closed with Gortex mesh.

In either type, investigation may reveal the presence of one or two portal triads and the anatomy of hepatic venous drainage. The degree of liver union is not predictable and the presence of a single portal vein or single common bile duct will pose significant operative difficulty. The plane of liver division may be obvious but pre-operative contrast enhanced CT scanning may provide a clearer indication of the plane of division. A manoeuvre which we have found useful is to develop a passage behind the two main masses of the liver and to use this bulk of liver tissue to digitally compress each half during the separation using an ultrasonic dissector and bipolar cautery to limit blood loss and seal vessels under vision. The raw surfaces of the divided liver are sealed with fibrin sealant.

Fusion of the foregut, often at duodenal level is common and the entire small bowel may be joined down to the site of a Meckel's diverticulum in the distal ileum.

Contrast studies have not been helpful in defining small bowel union and are not recommended. If the portal triads are separate, then each should be assured of sufficient intestine for survival. The presence of a single portal vein, single portal triad or single set of hepatic veins may preclude separation.

At the time of separation therefore, the availability of the ultrasound dissector and a vessel sealing device is essential to minimise blood loss during liver division.

Parapagus

The wide spectrum of parapagus union means that only a minority of these twins are candidates for separation. This may be possible if the hearts are separate.

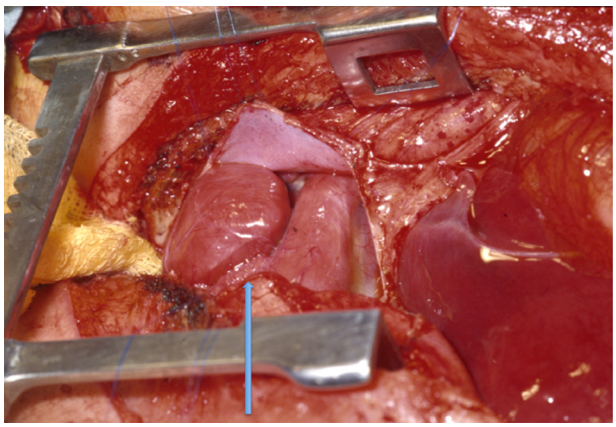

Fig. 2. Thoracopagus twins with separate hearts in a shared pericardial sac. The arrow shows the connexion at atrial level.

Liver union is dealt with as with other forms of union. Typically, the intestine is joined at the site of a Meckel's diverticulum and there is a single terminal ileum and colon. There may be one or two anuses and ano–rectal anomalies are common.

The urological anatomy is very variable. It is usually possible to determine the presence of one or two bladders, the level of union of the urethras and the ureteric anatomy prior to operation. Detailed investigation is worthwhile in order to facilitate decision-making during the operation. The presence of a single set of genitalia is also problematic and is dealt within the section on urology.

The final decision about who gets what is often the result of vagaries of blood supply and vascular anatomy. Decisions about allocation of bladder, urethra and genitals may be made prior to operation but are often altered by operative findings.

Ischiopagus

A wide spectrum of union is included in this group. The twins may be joined end to end with fusion of the two pelves and each twin having two lower limbs. Alternatively they may be joined at an angle where each has one normal lower limb and there is a third fused lower limb posteriorly.

The level of fusion in these twins may include the upper abdomen and the liver. Most often, in these twins, two portal triads are present.

The intestine is usually joined at the level of a Meckel's diverticulum and there is a single terminal ileum and large bowel. Occasionally, the caecum may be duplicated and there may be two appendices. Not infrequently, the single colon has a blood supply from each twin.

Ano–rectal anatomy is variable and ano–rectal abnormalities are frequent. A diverting colostomy is often needed in the neonatal period.

External genital anatomy is very variable. In some the external genitalia are duplicated, in others single. When there is a single penis, usually there are more than 3 corpora and there may be sufficient corpora to construct two penises. The testes are usually normal and may be descended. Some twins have one descended and one undescended testis. Ovaries are usually normal.

Internal anatomy is even more complicated and often asymmetric. Most have two bladders and two urethras. The manner in which these structures are allocated is quite individual and will depend on operative findings.

Union of the bony skeleton will depend on the variety of union. In the end to end variety, each has an open pelvis joined to the corresponding twin with two symphyses. In those with one normal leg each, a single anterior symphysis is present and the iliac bones are fused posteriorly. These twins have a third posterior dysplastic limb whose femur articulates with the abnormal posterior iliac bones.

Division of the liver and GI tract is performed as previously outlined. Allocation of the large bowel will depend on vascular anatomy

The details of genital and urological anatomy may be fairly clear prior to operation and the likely allocation of different structures tentatively planned. Prior cystoscopy and vaginoscopy would be considered mandatory. The place of laparoscopy is uncertain but easily performed if required. As much of the decision-making must await the time of separation, it is not necessary to undertake all possible investigations before this time.

A striking feature of separation of pelvic organs is the severity of haemorrhage which may be encountered.

When surgery takes place in infancy division of the bones or symphyses is not difficult and is associated with minimal blood loss. It is normally possible to remove the bones from the