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ARTICLE

Conjoined twins: an obstetrician's guide to prenatal care and delivery management

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OBJECTIVE: Obstetricians infrequently encounter conjoined twins. Much of the clinical care literature focuses on postnatal management from a neonatology and pediatric surgery perspective; guidance on obstetrical management is limited. We outline steps for prenatal evaluation, obstetrical care, and delivery planning.
STUDY DESIGN: Experiences with two cases of conjoined twins.
RESULTS: We identified several points throughout the planning, delivery, and postnatal process that are important to highlight for optimizing clinical outcome, patient safety, and parental satisfaction.
CONCLUSION: After diagnosis, patients should be referred to a center experienced in the management of conjoined twins. Specialists in fields including maternal fetal medicine, pediatric surgery, neonatology, and radiology play a vital role in the management of these patients. Early referral allows for timely family counseling and decision-making. Prenatal evaluation beyond the first trimester should include a detailed ultrasound, fetal echocardiogram, and fetal MRI. 3D printed life-sized models can improve delivery planning and patient understanding.

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INTRODUCTION

Conjoined twins are infrequently encountered by obstetricians. Many of these pregnancies do not achieve a viable gestational age due to miscarriage or pregnancy termination [1]. Much of the clinical care literature focuses on postnatal management from a neonatology and pediatric surgery perspective; guidance on obstetrical management of a conjoined twin pregnancy is limited [2, 3]. Our aim is to outline the steps for prenatal evaluation, obstetrical care, and delivery planning in a conjoined twin gestation.

METHODS

In this article, we share our experience with two cases of conjoined twins that continued to mid-third trimester and outline steps for prenatal evaluation, obstetrical care, and obstetrical delivery planning.

Background

Significant limitations exist within the epidemiologic data on conjoined twins, with the majority of reports being greater than 30 years old. Conjoined twins are rare, with an incidence of 1 in 100,000 to 1 in 250,000 live births, and are more common in non-Caucasian populations [4–6]. Best available estimates suggest that 1% of monozygotic twins are conjoined [1]. A predominance of female sex has been reported, although there is no biologic explanation for this [4, 6–8]. There is no evidence of an association with increased maternal age [4], and no genetic, environmental, or demographic factors are associated with the development of conjoined twins [6]. No good models for estimating the likelihood of livebirth for conjoined twins exist. A series published in 1982 [4] described a 60% livebirth rate, but standards and available technology for prenatal diagnosis differed greatly from contemporary prenatal care. The same series reported that 60% of live-born conjoined twins died within the first 24 h of life [4]. Thus, due to the high rates of stillbirth and neonatal demise, only 6–8 sets of conjoined twins survive to surgical separation each year [1].

Several theories exist behind the embryology of conjoined twins [9, 10]. The most commonly held hypotheses are the failure of the embryo to undergo complete separation between day 13 and 15 post-fertilization or the secondary union of two originally separate embryonic discs [9]. Multiple sites of connection are described, with both ventral or dorsal structures shared [9] (Supplementary Table 1). Of all possible fusion sites, the fetal chest is most common, making the majority of conjoined twins thoracopagus [6]. This suggests that the embryonic disc separates from both cranial and caudal ends, and if there is incomplete separation, the chest remains fused [6, 10].

Of conjoined twin types, thoracopagus also have the highest associated mortality rate due to the likelihood of shared vital cardiac structures [5]. Cardiac defects have been reported in up to 92% of conjoined twins, and extracardiac anomalies (e.g., limb, abdominal wall, facial defects) occurred in 62% of cases [11]. The reported stillbirth risk is likely related to the high incidence of anomalies, as well as strain on shared circulations [10].

Prenatal diagnosis

Routine prenatal ultrasound should equip the obstetrician with the ability to detect virtually all conjoined twin gestations. In the first trimester, a twin gestation with a single yolk sac or single placenta without a visible

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inter-twin membrane should prompt scrutiny of the fetal poles [1, 5]. Conjoined fetal poles may be intimately associated and lack independent motion. If the umbilical cord is visualized, it should be inspected for supernumerary vessels with a single placental cord insertion [5]. Some have reported that a definitive diagnosis can be made as early as 8–13 weeks' gestation [12–15]. Early diagnosis of conjoined twins provides the parents an opportunity for well-informed decision-making. Unfortunately, specific details about shared visceral involvement or cardiac structures that would impact counseling about long-term survival may not be possible until later in gestation. Although congenital anomalies are almost always present in conjoined twins and are not suggestive of a genetic etiology for the conjoined twinning, amniocentesis or chorionic villus sampling can be offered to exclude comorbid chromosomal abnormalities via chromosomal microarray [1, 6, 16].

For patients who did not undergo first trimester fetal ultrasonography, a detailed anatomic fetal survey at 18–20 weeks will be diagnostic (Supplementary Fig. 1). Complete sonographic evaluation to define the extent of fusion, shared viscera, and cardiac evaluation is essential for prognostic counseling. Due to the increased incidence of cardiac defects in monozygotic twinning, fetal echocardiography should be obtained regardless of the fusion site, as shared cardiac anatomy is associated with a poor prognosis [5]. Magnetic resonance imaging (MRI) may be a useful adjunct to clarify fetal anatomy that is incompletely defined sonographically, as well as to enhance details regarding brain, abdominal, and thoracic structures. This imaging should be performed in a center experienced in fetal MRI, as optimal scanning protocols and study interpretation are essential to future pregnancy and potential surgical management.

Prenatal assessment of surgical separation feasibility

Assessment begins with thorough prenatal imaging including ultrasound, fetal echocardiogram and fetal MRI to determine the anatomy of shared organs and associated anomalies. Based on these studies, the conjoined twins can be categorized into the proper anatomic subgroup according to the most prominent site of connection (thoracopagus, omphalopagus, parapagus, ischiopagus, pygopagus, and craniopagus), which will determine the multidisciplinary team required to make an assessment about feasibility of separation. Congenital anomalies are very common even in organs that are not shared and may impact mortality and morbidity risks for either twin independent of the feasibility of separation [4]. In the following paragraphs, we will review the factors that must be assessed to provide accurate prenatal assessment of the feasibility of separation. This is essential for counseling the family about all available options. It is important to discuss potential morbidity and mortality with separation so that the family can make an informed decision about the pregnancy.

Prenatal assessment may identify the need for emergent separation at birth, which predictably has a lower survival rate compared to planned separation within the first year of life [17]. Emergency surgery may be necessary if one twin is stillborn or has anomalies incompatible with survival, there is damage to the conjoined connection, or there are congenital anomalies that are surgically correctable but would be fatal if untreated [18]. These possibilities should be discussed with the family prenatally.

We will briefly delineate the general approach to prenatal assessment of surgical separation for the two most common types of conjoined twins. Thoracopagus twins are the most common, representing 40–60% of all conjoined twins [6]. Most have pericardial fusion, which can be separated. However, up to 75% of thoracopagus twins have conjoined hearts, which are usually inseparable [18, 19]. The severity of associated cardiac anatomic anomalies may also preclude successful separation. Prenatal echocardiogram assessment may be superior to postnatal examination because amniotic fluid acts as a buffer during ultrasound [18]. As such, fetal echocardiogram is essential to evaluate feasibility of separation which is often determined by cardiac anatomy and fusion. The liver is fused and the biliary tree is joined in 25% of thoracopagus cases [18]. Although fetal MRI is not definitive, it can identify separate gallbladders and separate hepatic drainage suggesting feasibility of hepatic separation without major reconstruction. Up to 50% of thoracopagus cases have connected gastrointestinal tracts which would require postnatal imaging studies to delineate. The ribs from each twin fuse with a shared sternum bilaterally. When this sternum is divided, each twin will have a narrow hemisternum which may be bridged with titanium plates to prevent a flail chest. However, chest wall mechanics cannot be entirely predicted. A shared diaphragm with fusion anteriorly is also common in thoracopagus twins, but can be separated [18, 19]. Complications such as prematurity and potential chronic lung disease will influence overall lung function after separation.

Omphalopagus twins are joined in the epigastrium and mid abdomen and account for ~30% of conjoined twins. The fused organs often include liver (80%) and intestine (30%) [20]. Although separation of omphalopagus twins is usually the least complex with favorable outcomes, morbidity will be determined by degree of liver and intestinal fusion. Fetal MRI may be helpful in identifying the degree of extrahepatic biliary tree fusion which may suggest the degree and location of intestinal fusion.

Ultimately, decision-making about surgical separation of the twins requires complex counseling—including whether a separation is felt to be feasible and safe; if there are discordant prognoses between the twins upon separation; optimal timing of separation procedure(s); and the complex psychosocial impact on the children and family. Neither a specific surgical separation plan nor an opinion that separation is impossible should be offered to the family without multidisciplinary input and coordination. The overall prognosis should be guarded in the prenatal period since the fetal natural history is not predictable and many factors influence outcome. For example, polyhydramnios is reported in up to 50% of conjoined twins and is a risk factor for premature delivery or rupture of membranes [21]. The risks of prematurity would be additive and could change the separation risk profile considerably. In general, timing of separation is at 6–12 months of life when the twins are bigger, tissue expansion can be performed to provide soft tissue coverage, and imaging and surgical preparation can be performed [22]. Clear communication between a multidisciplinary team and the parents is essential in this discussion. Table 1 highlights important considerations for the discussion with parents regarding surgical separation.

Prenatal care

For families faced with a conjoined twin diagnosis, obstetric counseling must explore the expectant parents' values while providing realistic and detailed expectations for the postnatal course. Counseling should start as soon as the diagnosis of conjoined twins is made. When counseling families on the diagnosis, it may be helpful to review images from the studies already obtained (such as the ultrasound or fetal MRI) and to provide diagrams of the fetal anatomy, as well as outcomes data. Options for pregnancy management including termination of pregnancy and expectant management should be thoroughly reviewed [23, 24]. The discussion surrounding expectant management may include perinatal hospice, palliative care, or a trial of therapy dependent upon parental wishes and expected postnatal prognosis [23, 24]. Regardless of whether or not a family desires postnatal separation, all

Table 1. Counseling on surgical separation.

Counseling Element	Details
What the discussion should entail	Is surgical separation feasible?
	What prognosis does separation impart for both infants?
	What prognosis does non-separation impart for both infants?
	Nature of surgical separation: • Nature and timing of initial procedure • Nature and timing of future procedures • Potential risks and benefits of the procedure for one or both infants (see above) and the short and long term
	Psychosocial and economic impacts
Who should be involved in counseling	Maternal fetal medicine
	Pediatric surgery
	Neonatology
	Pediatric surgical subspecialty applicable to type of conjoined twins (e.g., orthopedics, neurosurgery, urology, cardiovascular surgery)
	Plastic surgery
	Pediatric anesthesiology
	Palliative care
	Social work

patients should be counseled about all the potential outcomes as specifically as possible, including long-term morbidity and mortality risks, with acknowledgement of the limited data to inform this counseling [25]. Anticipatory guidance on the nature of possible interventions, likely events in the neonatal intensive care unit (NICU), and estimated length of NICU stay should be provided; coordinated consultation with a neonatologist and other pediatric subspecialists about postnatal care is advised. Careful attention to care coordination around multiple subspecialty consultations tailored to the individual family's needs and preferences is essential for ensuring that families are counseled consistently and do not receive contradictory information [26]. The potential benefits of integrated multidisciplinary consultation should be balanced with the risks of overwhelming the expectant parents with large panels of subspecialists, but consistency with one or more providers throughout serial consultations may be helpful in providing continuity and clarification. Throughout this process, patients should be provided with resources including support groups and therapists for the psychosocial stressors and family issues that may arise from such a complicated pregnancy [27]. A focus on family support and a discussion regarding media exposure to prepare families and providers should also occur [27]. Finally, for patients who choose to continue the pregnancy, relocation may be needed to accommodate the extensive prenatal needs of the mother and postnatal needs of the infant [27]. If relocation is undertaken, it should occur at the time fetal intervention would be considered.

Termination of pregnancy. Based on data from a single center study, 50–70% of patients elect termination of pregnancy after comprehensive consultation [11]. Termination care is gestational-age dependent with dilation and curettage (D&C), dilation and evacuation (D&E), hysterotomy, and labor induction all described [14, 28]. The most appropriate management minimizes maternal risk and considers the site of connection and gestational age. Local laws must also be considered when counseling patients, as legal restrictions related to termination limits vary. In addition, costs of termination and anesthesia considerations with each method of termination should be made available to patients, as this may impact their decision-making process.

Non-surgical termination: As is true with all pregnancies, earlier termination minimizes the risk of maternal morbidity [1]. Although vaginal delivery is not recommended for conjoined twins at or near term, at earlier gestations, fetal pliability may make an induction termination a safe and feasible option. Pre-procedure counseling for an induction termination should include potential risk for labor dystocia, hemorrhage, and need for hysterotomy or urgent D&E if these complications occur. Clinicians should account for fetal presentation, widest diameter of the fetuses, and fusion type. Although induction termination at later gestation may pose increased maternal risk, termination in the late second trimester via induction may be possible. Successful vaginal births up to 25-weeks' gestation of thoraco-omphalopagus and pyopagus conjoined twins has been reported [29]. If induction termination is elected, clinicians should discuss the utility of induced fetal demise prior to the induction, as well as use of laminaria, mifepristone, and misoprostol—all of which can be used to promote cervical effacement and dilation [29]. Inductions should occur in institutions with the capability to manage major labor or surgical complications. Undergoing a successful vaginal delivery for induction allows the patient to avoid surgery (e.g., laparotomy or D&E). The possibility for the family to see and hold the fetuses afforded by vaginal delivery may inform their decision-making. However, this approach may not be appropriate in all cases and we underscore care individualization.

Surgical termination: Gestational age and availability of clinicians trained in surgical termination techniques will affect available options among D&C, D&E, or hysterotomy. Although hysterotomy has historically been employed for surgical termination, we recommend against this approach given the associated maternal morbidity [28, 30]. Even in the second trimester, D&E by an experienced provider is the first-line surgical option, as the associated morbidity is significantly less than with hysterotomy. If D&C or D&E is selected, assessment of fetal size, fetal presentation, fetal width, and fusion site to determine the extent of indicated cervical preparation (i.e., number of millimeters of laminaria) and use of adjuvant medications including mifepristone or misoprostol should be made. Similar to induction termination of conjoined twins, surgical termination should be performed in a center with the capability to manage major surgical complications.

Table 2 provides an overview of the risks and benefits with each termination method. If the patient is in the first trimester, we support D&C as the safest option for pregnancy termination. If the patient is in the second trimester, decision-making should be individualized for each patient based on a number of factors including any potential maternal comorbidities, a family's personal values, fetal presentation, fusion type, and fetal size. Although there are no studies comparing risks of D&E versus labor induction termination for conjoined twins, a recent review of second trimester pregnancy termination for fetal indications demonstrated no difference in risks between either method [31].

Continuation of pregnancy. Available literature offers no clear guidance on obstetric surveillance for patients pursuing expectant management for conjoined twins. Given this lack of specific evidence-based guidance, we recommend balancing the values and preferences of the expectant parents and best-available evidence for monitoring of monochorionic pregnancies [32]. The clinician should assess the expectant parents' values when creating the care plan, and consideration should be given to what pregnancy outcomes are acceptable for them.

Based on underlying monochorionicity and higher rates of reported stillbirth, for patients desiring neonatal intervention, fetal assessment via sonography should be completed at least monthly. The fetuses should be monitored for growth, amniotic fluid volume, and signs of cardiac failure, as well as further definition of anatomy. Doppler measures typically assessed in monochorionic twinning, including umbilical artery, middle cerebral artery, and ductus venosus evaluation, may be more challenging to interpret due to circulatory anastomoses and fusion sites. Nonetheless, if there is evidence of growth restriction of one or both fetuses, we recommend obtaining umbilical artery Doppler evaluation for each fetus on all umbilical arteries. If these are abnormal, consideration may be given to altering timing of delivery. If no growth abnormality is detected, we recommend initiation of twice weekly antenatal testing whenever intervention would be considered—usually at 32 weeks. A biophysical profile may be required due to difficulty of obtaining two distinct fetal heart tracings, depending on the site of fusion. If interval growth and Doppler evaluation or antenatal testing are not reassuring, betamethasone administration and acceleration of delivery planning should be discussed and balanced against risk of stillbirth with ongoing expectant management. This should all be informed by the postnatal care plan and expectant parents' values. We recommend delivery at 34–36 weeks' gestation to balance the cumulative risks of stillbirth versus complications of prematurity, as extrapolated from care of nonconjoined monochorionic twins. Unless the patient has chosen to pursue a comfort care only approach, antenatal betamethasone should be administered prior to delivery.

As with any pregnancy, there is the potential risk that the twins may deliver (due to iatrogenic indications or spontaneously) prior to a gestational age at which intervention may be feasible. Such situations

Table 2. Overview of risks and benefits with each termination type.

Procedure	Risks	Benefits
Non-surgical termination • Labor induction	• Failure of induction, possible prolonged length of stay • Need for emergent surgery • Injury to the cervix	• Parents may see and hold the fetuses • Availability of complete postmortem anatomical exam
Surgical termination[a] • Dilation and curettage (D&C) • Dilation and evacuation (D&E)	• Typical surgery risks (e.g., bleeding, infection, thrombotic events) • Need for repeat procedures • Injury to the cervix	• Ability to avoid process of labor • Quick, outpatient procedure

[a]Would not consider a laparotomy/hysterotomy as a standard option.

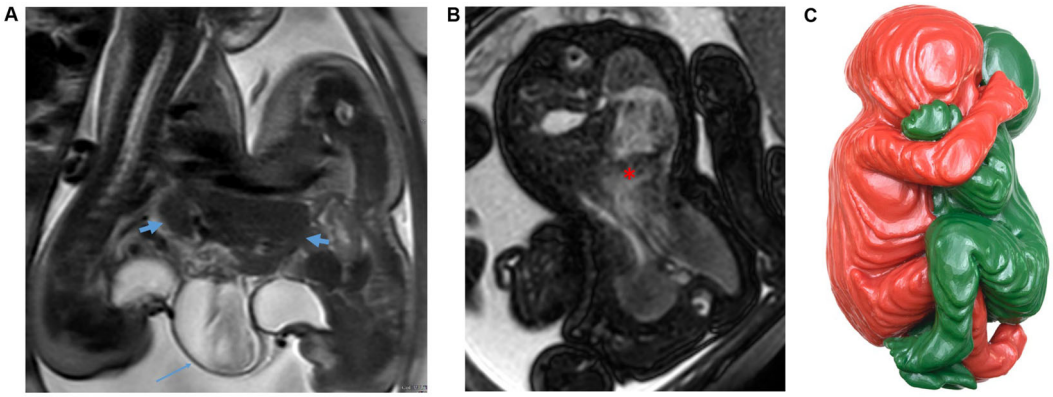

Fig. 1 Fetal MRI, case 1. A. Sagittal T2 weighted SSTSE (single shot turbo spin echo). **B**. Axial balanced FFE (fast filled echo). Conjoined twins with "shared" heart (*) primarily committed to twin A (to the right and to the top) and elongated cardiac chambers with shared ventricle located inferiorly along the diaphragm. Conjoined liver (short arrows) primarily committed to twin A and shared small bowel, with a dilated fluid-filled bowel loop, which protrudes inferiorly through an abdominal wall defect/omphalocele in the inferior shared abdomen (long arrow). **C**. 3D printed model. The 3D model shows each of the twins facing each other with arms and legs wrapped around each other. Model was printed solid and weighted 4.24 kg, similar to the actual delivered weight of the twins.

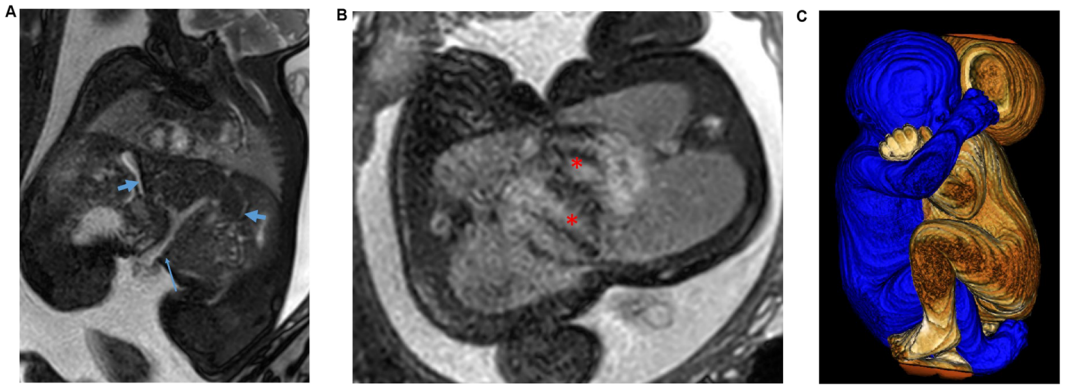

Fig. 2 Fetal MRI, Case 2. A. Sagittal T2 weighted SSTSE (single shot turbo spin echo). **B**. Axial balanced FFE (fast filled echo). Conjoined twins with two abutting separate hearts (*), each heart with four chambers and apex oriented to the left (relative to each twin). Conjoined liver with slightly larger portion committed to twin A. In each twin, the liver is predominantly within the right upper quadrant (short arrows). A single umbilical vein (long arrow) enters the abdomen inferior to the conjoined liver with early bifurcation and connection to the corresponding portal venous system for each twin. **C**. Volumetric 3D rendering.

need to be approached on an individualized basis, taking into account the current gestational age, anomalies present, and site of fusion. Based on the information available, early counseling should include the limitations in care that would be posed by prematurity. These discussions should not wait for an urgent situation, although scenarios may require reassessment with core members of the care team (maternal fetal medicine, neonatology, and pediatric surgery) regarding feasibility of postnatal management and interventions at that point in time.

Even if a previous MRI was performed, a third trimester MRI is recommended if there is a plan for possible postnatal interventions. This will allow better delineation of fetal visceral structures, assessment of fetal size and relationships, and facilitation of delivery planning. 3D modeling with subsequent 3D printing can be performed as an adjunct to assist with delivery planning, including size and location of the uterine incision (Supplementary Fig. 2) and detailed planning of neonatal resuscitation.

Delivery management

The diagnosis of twins itself has implications that put the pregnancy at high risk of morbidity. A prenatal diagnosis of conjoined twins further influences timing, location, and mode of delivery.

Maternal risks of multifetal gestation include a higher incidence of hyperemesis gravidarum, hypertensive disorders, anemia, and gestational diabetes. Postpartum, the risk of hemorrhage is increased due to the uterine overdistention of a multifetal pregnancy. Preterm birth risk is increased due to both maternal (e.g., pre-eclampsia, preterm labor) and fetal (e.g., polyhydramnios, growth restriction) etiologies [1]. Expanded maternal risk also extends to delivery—the majority of conjoined twins are at risk of obstructed labor. If the fetuses are unable to pass through the maternal pelvis and labor is allowed to continue, there is a risk of uterine rupture. If cesarean is undertaken after prolonged obstructed labor, there is a higher risk of maternal morbidity including postpartum hemorrhage or damage to the cervix [1]. For conjoined twins in the third trimester, the recommended mode of delivery is cesarean, regardless of the plans for neonatal care. Cesarean delivery decreases the risks of avulsion injury to shared fetal tissues, related risk of internal hemorrhage, and intrapartum demise. Skin and uterine incision types must be carefully considered. Traditionally, a vertical skin incision with a vertical uterine incision (classical cesarean delivery) has been recommended. However, in certain scenarios, a Pfannenstiel skin incision followed by either classical or low transverse incision on the uterus may be considered based on dimensions of the combined fetal mass, anatomy of fetal attachments, fetal presentations, placental location, and maternal and uterine anatomy [1].

In our experience, a third trimester life-sized 3D printed model from MRI was very helpful for delivery planning (Figs. 1 and 2). Exact sizes and anatomical relationships allowed individualized planning for the uterine incision. A 3D printed model that has a similar weight to the fetuses functions well for simulations, as it allows the delivery teams to develop a sense of the expected weight of the conjoined twin gestation versus a nonconjoined twin gestation at birth. The model is also helpful for the neonatal team in planning resuscitation efforts, including transfer of the neonates from the operating table and positioning of the neonates for post-delivery management. Knowing the position of the airways is helpful to optimize positioning for intubation. The 3D printed model also yielded visual aids specific to each conjoined pair for family counseling

regarding surgical morbidity to support shared decision-making for each fetus.

Safe and controlled delivery and resuscitation of conjoined twins requires extensive planning. This includes careful attention to delivery and resuscitation teams and exploration of available space and resources for such a complex and rare event. Supplementary Fig. 3 provides an example layout of the operating room and neonatal resuscitation area. We recommend two main teams: an obstetrical team, headed by a maternal fetal medicine specialist along with at least one additional obstetrician, and a neonatology team, headed by a neonatology specialist with two sub-teams—one for each infant.

Resources should be available for each infant to have a separately designated complete resuscitation team, in accordance with Neonatal Resuscitation Program guidelines [33]. Modifications to the Neonatal Resuscitation Program guidelines may also need to be considered based on the anomalies that have been identified in the twins prenatally [34]. Clear delineation of responsibilities for everyone in the delivery room/resuscitation area are necessary, and careful, prospective attention to the balance of crowd control and educational opportunities is required. It is also important to have a contingency plan for airway management, including consultation with pediatric anesthesia and/or otolaryngology in advance of the delivery to determine necessary personnel and equipment for airway management at delivery. Which specialists should be present to participate in the care of the infants immediately after birth will depend on the individual anatomy and physiology and the previously determined care plan, with contingency planning for anticipated complications.

Simulation of the delivery and resuscitation, with time for revision and repeated planning exercises should be undertaken well in advance of the delivery given the complexities of care coordination and obstetric and postnatal care, as well as the risk for urgent or emergent preterm delivery. All specialists expected to participate in the perinatal care of the conjoined twin pair should be included, ideally with the same individuals expected to be present for the delivery. One or more simulations are vital to rehearse roles and steps in postnatal management of the infants to streamline their care and improve stabilization outcomes [1]. There are multiple publications that review simulation-based training in preparation of conjoined twins [2, 3, 35].

We recommend at least two obstetricians perform the delivery to assist in minimizing fusion-site injury. Immediately prior to the delivery, verification of the positioning of the fetuses with ultrasound is helpful to confirm identification of each fetus for facilitation of individualized care. Immediately after delivery, while still on the operative field, we recommend labeling each infant so prenatal information is accurately assigned to guide postnatal care. Post-delivery labeling can be done by putting a sterile hat on each infant while still on the surgical field (Supplementary Fig. 4). We recommend immediate cord clamping in monochorionic twin gestations due to shared circulations. Following cord ligation, the infants can be handed off for assessment and stabilization by the neonatologists [1].

Finally, an ex-utero intrapartum treatment (EXIT) procedure may be considered in certain high-risk neonatal deliveries. An EXIT should not be the routine approach for delivery of conjoined twins, as the procedure has high maternal morbidity and may prevent the mother from seeing the neonates if there is early neonatal demise. It should only be considered in a delivery if there is prenatal imaging evidence of airway compression in a twin and if obtaining an airway on the delivery field is realistic and has the potential to improve outcomes for the neonates.

RESULTS

To illustrate our recommendations, we offer a review of two recent patients who received care at our institution. Case 1 presented for initial consultation in April 2019 and was delivered in April 2019. Case 2 presented for initial consultation in February 2019 and was delivered in June 2019. Both patients gave signed consent to participate in this review.

Case 1

A 23-year-old G2P1001 female presented at 34 4/7 weeks' gestation for third opinion consultation in a pregnancy complicated by thoraco-omphalopagus conjoined female twins. The diagnosis was made in the first trimester, at 11 weeks' gestation. She had a non-contributory medical history and one prior term vaginal delivery. She had received care at an outside facility with a plan for delivery via cesarean section near term with perinatal hospice for the infants. She and her partner were interested in further evaluation and counseling about the possibility for postnatal intervention; specifically, if there was any potential for long-term survival for at least one of the fetuses if surgical separation were attempted.

Evaluation included detailed ultrasound, fetal echocardiogram, fetal MRI, and multidisciplinary consultations involving maternal fetal medicine, pediatric cardiology, pediatric surgery, neonatology, pediatric radiology, palliative care, and social work. The parents declined consultation with spiritual care. Ultrasound findings included confirmation of thoraco-omphalopagus conjoined twins in vertex presentation with positional deformity of the skull for fetus A; kyphosis of the spine in fetus B; a shared liver, with the majority situated in fetus A; an umbilical cord with five vessels; possible ventral herniation of the bowel near the umbilical cord insertion site; and shared complex cardiac anatomy. Fetal echocardiogram demonstrated the cardiac position primarily in the chest of fetus A with three ventricles, three sets of atrioventricular valves, and preserved systolic function in each ventricle. In addition to confirmation of the above findings, MRI demonstrated low lung volumes for each fetus (30% of expected in fetus A and 20% of expected in fetus B) and conjoined bowel with abnormally dilated loops suggestive of bowel atresia. Ultimately, the parents elected delivery and a trial of therapy. Given the combined fetal width and presentations, the parents were counseled on the need for a vertical skin incision and vertical uterine incision.

Using a 3D modeling of the fetuses rendered from MRI imaging, a team-based approach—including the obstetricians, neonatologists, pediatric cardiologists, pediatric surgeons, and nurses involved in the patient's care—was utilized in delivery planning of the fetuses (Fig. 1C).

After a course of betamethasone, the patient underwent scheduled classical cesarean section at 35 5/7 weeks. Pre-delivery ultrasound confirmed fetal vertex positioning and placental location. Following vertical skin incision, an anterior vertical uterine incision was carried down to the level of the amniotic sac. Both fetal heads were guided towards the incision prior to performing the amniotomy. Once the amniotomy was performed, both heads delivered simultaneously. This was followed by elevation of the united bodies by both delivery obstetricians, with care to avoid traction forces that may avulse shared tissues (Supplementary Fig. 4). The quantitative blood loss was 1680 ml, which the mother tolerated without transfusion. She did well postoperatively and was discharged on postoperative day #2.

Due to continued apnea and poor oxygenation despite increasing PEEP and FiO2 requirements with CPAP, the neonates required intubation shortly after birth. They underwent extensive evaluation with echocardiography, CT angiography, and renal and transcranial sonography. Cardiac dysfunction developed in the postnatal period, and a multidisciplinary evaluation determined the complexity of the cardiac function and anatomy was incompatible with long-term survival for either twin, with or without separation. Transition to comfort care was made, and both infants died on day of life #15.

Case 2

A 32-year-old G2P1001 female was referred at 19 3/7 weeks' gestation for consultation after an anatomic survey at an outside facility demonstrated a conjoined female twin pregnancy that was undiagnosed at the 8-week ultrasound. She had a non-contributory medical history and one prior term vaginal delivery. The ultrasound findings included thoraco-omphalopagus conjoined twins with fusion of the chest wall beginning at the level of the sternal notch and extending below the ventral cord insertion

Table 3. Key learning points from our experiences.

Time Frame	Learning Points
Prenatal	• The simplicity and consistency of integrated subspecialty counseling should be balanced with parental emotional and learning needs to make an individualized plan for information sharing. • Available hospital resources should be explored and leveraged to optimize outcomes. • Creating a 3D model of the fetuses can greatly enhance delivery planning and optimization. • Delivery and resuscitation simulation with the members of the planned care teams is essential given that conjoined twin birth is a rare event and institutional experience is limited.
Day of Delivery	• Clinical continuity with physician service leads is essential for seamless and safe patient care for a highly complex and choreographed rare event. • Clearly designated leaders are essential to prospective plans for crowd control, chaos reduction, and patient privacy.
Postnatal	• The extraordinary conditions of a conjoined twin delivery should not eclipse the importance of routine measures to ensure compassionate family centered care.

(Supplementary Fig. 1). There was no identifiable sternum. There were two 4-chamber heart structures with abutting walls of the ventricles. Each fetus has a liver with separate gallbladders but the liver was fused in the midline. An inferior vena cava was visualized only in fetus A. No organ anomalies independent of the conjoined structures were identified.

Following the ultrasound, the patient and her partner received extensive counseling by the maternal fetal medicine specialist with discussion of the findings, potential implications, and options for the pregnancy. To support decision-making, she underwent a fetal MRI and fetal echocardiogram at the referral institution. The MRI confirmed the ultrasound findings of a single umbilical cord comprised of a single umbilical vein entering the abdomen inferior to the shared liver and bifurcating into two separate portal venous systems (Fig. 2A). The fetal echocardiogram confirmed two separate hearts with normal intracardiac anatomy.

The couple had additional consultations with pediatric surgery, pediatric cardiology, neonatology, palliative care, pediatric radiology, social work, and spiritual care about the findings, potential outcomes, and options. There was extensive discussion about chest well reconstruction that would accompany any surgical separation. For this couple, the possibility of surgical separation with survival of one or both of the twins informed their decision of pregnancy continuation. The couple was counseled on the goal of delivery at 34–36 weeks' gestation, possibly via classical cesarean section. The plan was made to continue with monthly obstetrical visits and ultrasounds until 32 weeks, at which time weekly biophysical profiles were initiated. At 33 weeks, the patient underwent a second fetal MRI in order to assist with delivery planning. Concurrently, an umbilical artery Doppler study revealed new onset absent end diastolic flow. The decision was made to proceed with planned delivery at 34 weeks after administration of betamethasone.

Using images from her third trimester fetal MRI (Fig. 2), a life-sized 3D printed model of the fetuses was made. Similar to Case 1, a simulation of the delivery and neonatal resuscitation was performed using the 3D printed model.

On the day of surgery, a pre-delivery ultrasound was performed to determine placental location and fetal positions. Both infants were breech and the placenta was anterior. The skin was marked for a 22 cm incision based on the 3D print of the fetuses. At the level of the uterus, sterile intraoperative ultrasound was performed to map out the optimal uterine incision location relative to the placenta, as well as the needed size for the incision (Supplementary Figure 2). A low transverse incision was made on the uterus with extension into a "J" fashion on the left-hand side. The breech fetuses were delivered by grasping all four legs and providing gentle traction, while the second obstetrician made sure the upper extremities delivered freely (Supplementary Figure 5). The uterine incision remained in the lower uterine segment and provided adequate room for a safe delivery. The mother did well postoperatively and was discharged on postoperative day #2.

The neonates were placed on CPAP and stabilized immediately after birth. In the weeks following their birth, they underwent extensive imaging. The infants did require intubation during their NICU course, but were ultimately weaned to nasal cannula prior to discharge. They were discharged to home on day of life #85, with plan for separation prior to one year of life. Postnatal 3D modeling has also been utilized in the planning of their upcoming separation (Supplementary Fig. 6).

DISCUSSION

Review of these cases identifies several points throughout the planning of prenatal, delivery, and postnatal care that are important to highlight in order to optimize clinical outcome, patient safety, and parental satisfaction. Table 3 summarizes these points.

Following the referral of the first patient, a need for rapid consultation with multiple services was required due to advanced gestational age. The teams were assembled quickly, but the limited time for discussion posed challenges. To ensure consistent communication and planning in such a short pre-delivery window, a multidisciplinary group meeting was held with the family; this experience was quite overwhelming for the couple. For the second couple, spacing out individual meetings with the various providers over several weeks allowed easier comprehension of the complex diagnoses and plans of care. It is important that all information is obtained and shared with the patient within a timeline that will allow for decision-making to include the option of termination of pregnancy. For couples continuing the pregnancy, we believe a team-based approach, with essential prenatal consultations spaced out over time, optimizes assimilation of complex information and allows for adequate communication amongst family and providers to create a shared agreement on the delivery and postnatal management plans.

Next, while consultations and evaluations are ongoing, we recommend consultation with hospital legal and media teams for guidance on how best to protect patient privacy. As with all patient information, sharing of information should be on a "need to know basis." Some families take comfort in sharing their journeys with a difficult pregnancy or infant care, while others are more private. A recent publication underscores a focus on family support and their desire for media exposure to prepare families and providers when rare cases such as conjoined twins are delivered [32].

From a delivery planning standpoint, late third trimester MRI-based 3D printing of the fetuses to display positioning and size was extremely helpful (Fig. 1C). The 3D model was printed full-size in plastic, with each fetus printed in a separate color. It was then utilized in pre-delivery simulation of the delivery, hand-off, and positioning of the fetuses during resuscitation, and also allowed individualization of the maternal abdominal and skin incisions. Creating a 3D printed model of the fetuses optimizes delivery planning and care.

Medical record numbers were created, and medication orders were placed, for each anticipated infant one day prior to delivery to minimize potential administration delays. Dosages were based on recent sonographic weight estimates. On the day of delivery, a brief preoperative meeting was held before the mother was brought back to the operating room. This verbal "time-out" confirmed roles, including the lead physician making decisions for the delivery, the individual who would transfer the neonates to the resuscitation bed, the neonatologists responsible for each neonate (sub-teams), and the lead neonatologist who would make final decisions about continuation or cessation of resuscitation efforts. Additional roles included an individual to provide communication and support for the patient and her partner throughout the delivery and initial infant care and individual(s) to document in each separate medical record. Once the patient was in the operating room and positioned, the roles of involved members were again verbally confirmed and an ultrasound was performed to confirm twin A versus twin B positioning to eliminate potential confusion when applying prenatal findings to postnatal care. As not all providers may be able to attend the pre-delivery simulation(s), it is imperative to designate lead providers to handle delivery day questions and coordination.

During the birth of the twins, personnel were designated to prevent unnecessary traffic in the areas outside of the operating room and resuscitation areas. Retrospectively, additional signage or privacy screens may have been helpful in specifically directing unnecessary traffic away from the resuscitation area for the first case. This was improved for the second case and effectively decreased traffic. Having both designated personnel and posted signage outside the operating rooms to prevent unnecessary traffic is essential for patient privacy and safety while allowing adequate space for the large multidisciplinary care team.

Following the deliveries, the father was able to move freely between the resuscitation area and the mother, who remained in the operating room. He was able to trim the umbilical cord, allowing for a bonding moment with his daughters. After completion of the cesarean section, and prior to maternal transport to the recovery room, the mother and father in both cases were relocated to the neonatal resuscitation area for a period of quiet time to facilitate bonding. This was important regardless of whether hospice care or resuscitation was planned [27]. Facilitating protected time with the infants early in their clinical course is essential in family-neonate bonding and coping with complex deliveries, neonatal intensive care, and grief.

Summary

Conjoined twin gestation is a rare event, which has unique complexity for obstetric management regardless of the patient's goals of care. Provision of coordinated, expert, and compassionate care requires extensive mobilization of resources for delivery and postnatal care, with iterative steps of prognostication and medical and surgical care planning for patients wishing to carry their pregnancies to near term. Fundamental principles of sound obstetric care and family-centered postnatal care should be applied, but clinicians may face additional obstacles related to the complex medical needs of conjoined twins. Once a diagnosis of conjoined twins is made, the patient should be referred to a comprehensive fetal therapy center with the resources and expertize for extensive prenatal evaluation, prognostication, and treatment. Early referral, when possible, allows for timely counseling and exploration of patients' values. They should be counseled by specialists in at least the following fields: maternal fetal medicine, pediatric surgery, neonatology, palliative care, and social work. Pediatric cardiology consultation should be considered in all fusion types, as cardiac function can be impacted by shared circulations. Early referral with comprehensive counseling allows adequate time for patients to assimilate complex information and understand decision-making implications. Depending upon the twin fusion site, consultation with additional services may be appropriate (e.g., neurosurgery for cephalopagus twins, cardiothoracic surgery for thoracopagus twins). Prenatal evaluation beyond the first trimester should include at least a detailed ultrasound, fetal echocardiogram, and fetal MRI. For patients who choose expectant management, ongoing evaluation to follow fetal growth and well-being, as well as an iterative assessment of feasibility of separation, may be used for further counseling and decisions regarding delivery timing. We found 3D models to be very helpful for educating families and planning uterine incision and neonatal resuscitation details. A fully orchestrated, team-based prenatal and delivery plan will improve the quality and safety of patient care.

DATA AVAILABILITY

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

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AUTHOR CONTRIBUTIONS

PG formulated the concept of the paper, wrote the entire manuscript, researched prior publications, and corresponded with the co-authors on their input. DP provided input on the manuscript. WW and ML created the images and models used in the described cases and provided input on the manuscript. NL provided input on the manuscript. GM provided input on the manuscript. AC helped with formulation of the concept behind the paper, researched prior publications and provided input on the manuscript. MT helped with formulation of the concept behind the paper, researched prior publications and provided input on the manuscript. All authors approved the final submitted version of the manuscript and agree to be accountable for all aspects of the work.

CONFLICT OF INTEREST

The authors declare no competing interests.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

This study was performed in accordance with the Declaration of Helsinki. Both patients signed consent to participate in this review.

ADDITIONAL INFORMATION

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