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An ethically-justifiable, practical approach to decision-making surrounding conjoined-twin separation

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Abstract

Conjoined twins present unique ethical and palliative care challenges. We present an ethically-justifiable, practical approach to decision-making with regards to surgical separation. These decisions must account for the short- and long-term prognoses for each infant prior to, and after, separation. Other considerations include the benefits and burdens of separation and the family's values and goals. Caregivers should recognize that decisions surrounding separation may be unduly influenced by social biases.

The palliative care team aids in developing goals of care to guide decision-making by promoting communication between the medical team and family. They play an important role in supporting families regardless of the planned course of treatment. This support may be social or spiritual in nature, and is promoted by the interdisciplinary structure of the team. Early involvement of palliative care services facilitates complex decision making and can aid in the transition from cure-oriented therapies to support if needed during and after the dying process.

Keywords

Palliative care; Ethics; Neonatology; Conjoined twins; Complex decision making; Interdisciplinary team

Introduction

Due to the high risk of severe congenital anomalies, conjoined twins often present with potentially life-limiting conditions. As such, complex decision-making and care at the end-of-life often play an important part in their management. Due to their status as conjoined persons, decision-making and palliative care must account for the challenges posed by this unique condition. We first present an ethically-justifiable, practical approach to decision-making regarding surgical separation taking into account the prognoses of the infants prior to and after separation, the benefits and burdens of separation, and values and goals of the family. We then move to considerations in the provision of palliative care for this group of infants.

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A practical approach to decision-making

The ethics surrounding the separation of conjoined twins are complicated. Risking, or even sacrificing, the life of one twin to save the other is a source of contention. Societal pressures to achieve “normality” add a psychosocial domain to be considered. Ultimately, these patients force us to question our understanding about the nature of individuality and what it means to be a “person.” Many authors have opined on the philosophical debate surrounding the separation of conjoined twins. Instead of contributing to the aforementioned debated, we instead will focus on an ethically-justifiable, practical approach to decision-making surrounding separation.

In Table 1, we have grouped clinical scenarios by predicted outcomes both prior to and after separation. We separated survivors into those expected to survive less than 6 months, and those expected to survive for longer. This distinction is not arbitrary. The appropriate care of patients expected to survive less than 6 months is end-of-life or hospice care.¹ As such, the care of these patients should focus not on surgical separation and post-surgical management, but on the alleviation of burdensome symptoms, spending quality time with the family, and honoring the dignity of the child and their family.¹ Nonetheless, the 6 month time frame should not be considered a hard and fast rule. If survival is expected to last beyond 6 months, but that time is likely to be dominated by heavy iatrogenic burdens and little opportunity to enjoy life, then such patients should be categorized with those expected to die before 6 months.

When both twins die if remain conjoined

In the event that the twins will die if they remain conjoined, but will die within 6 months if separated, it is impermissible to attempt separation. The burden of separation is too great for any expected benefit stemming from separation. If survival of both twins is expected to exceed 6 months, then it may be permissible to offer separation to the family. It is important however, to temper surgical enthusiasm for attempted separation with a sober assessment of the projected benefit. Decision-making should consider the family’s values and goals of care in the context of the benefits and burdens of post-surgical and long-term management.

Separation may be permissible or even obligatory if one twin is expected to survive long-term. The degree of long-term morbidity should be the deciding factor on whether separation should be merely permissible, or be considered obligatory. However, there are limitations. Because conjoined twins often share organs, it may be impossible to provide both twins with sufficient functioning organ tissue to survive. For example, thoracopagus twins often share cardiac tissue and have circulatory systems which are functionally inseparable,² such as in the case of the Maltese twins, Jodie and Mary.³ In such scenarios, any attempt at separation must therefore sacrifice one twin to save the other by separating the heart from said twin. Such a procedure is ethically impermissible if it requires that one twin be sacrificed to salvage the life of the other.

Justification for this limitation rests on three considerations. The principle (or doctrine) of double effect (see Table 2) has four conditions, which must be met for an action to be

morally permissible. In this scenario, the act of removing the heart from one twin cannot be considered a morally indifferent act. This should be obvious, as we cannot justify any other scenario in which removing the still functioning heart from one patient, even one near death, for the benefit of another would be condoned. This act also fails the second condition in that the bad effect, removing the conjoined heart to which the sacrificial twin has at least a partial, if not equal claim is the means by which the other twin can be salvaged. A previous analysis of the case of conjoined twins using the principle of double effect supports this approach.⁴

The second justification is based on patient preference. This approach may seem counterintuitive as the traditional approach to decision-making for infants is based on best interests.⁵ However, conjoined twins belong to a unique population whose interests are difficult to comprehend for anyone not born into it. While infants cannot express their preference for surgery, we can look at the expressed wishes of patients in similar situations for guidance. When asked about whether or not they desired surgical separation if it meant that their twin might die, or even to save their life after their twin died, conjoined twins expressed a strong preference for remaining conjoined.⁶ These data argue against sacrificing the life of one twin for the benefit of the other based on a calculation of best interests done by anyone other than the twins themselves.

Analogies in transplant ethics provide the third justification. The AAP has published conditions which must be met in order for a minor to donate a solid-organ.⁷ One of the conditions is that both donor and recipient must be certain to benefit. Since the donor cannot physically benefit from donating an organ, there must be a psychosocial benefit to be gained by the donor. This benefit allows for a twin to accept some risk of morbidity for the benefit of his or her sibling. However, other conditions require that the surgical risk for the donor be extremely low, and that the donor be able to assent to the procedure. For the medical team and parents to decide that one twin must sacrifice not only an organ, but also his or her life for their sibling violates all three conditions. The AAP makes no allowance for donation if the donor is near death. Thus, while it may be permissible to submit a dying twin to the risk of death for the benefit of their sibling, it is impermissible to cause their death for that same end.

If both twins are expected to survive the separation surgery, but either one or both are expected to suffer from considerable morbidity, surgical separation is permissible. Once again, decision-making should consider the family's values and goals of care in the context of the benefits and burdens of post-surgical and long-term management and the expected quality of life for the infants. If no major morbidity is expected for either twin, then surgical separation should be considered obligatory.

A scenario which generates a great deal of debate is what to do when the death of one twin threatens the life of the other. If the surviving twin is stable and thought to benefit from separation, then separation should proceed. Organs and tissues that are shared, or that are thought to "belong" to the deceased twin, should be considered available to be used as needed for the benefit of the survivor, analogous to those of a deceased organ donor.

More contentious is the course of action if the likelihood of survival of one twin is improved if separation occurs before the doomed twin dies. Again, it is not permissible to sacrifice one twin to benefit the other, therefore surgical separation with the intent of hastening the death of one (for example, by harvesting its organs or tissues) to benefit the other is impermissible. As discussed above, it may be permissible to subject the doomed twin to surgical risk if done without intention of hastening death and if it clearly benefits the expected survivor. This is a difficult moral quandary and such decisions would benefit from a multi-disciplinary approach including the palliative care team and involvement of the ethics committee.

When both twins survive if remain conjoined

Surgical separation is impermissible if one or both twins are expected to die from the procedure when they otherwise would have remained alive conjoined. No expected benefit for a potential survivor can outweigh the other twin's loss of life. If both are thought to survive separation surgery and one or both are expected to suffer from severe post-surgical morbidity, then separation may be permissible. Considerations for decision-making should proceed as before, but extra weight should be given to whether the calculation of best interests and quality of life are being made from the perspective of the twins, or others struggling to appreciate their unique perspective. If neither is expected to suffer from long-term morbidities post-separation, then the procedure should be considered permissible, and potentially even obligatory.

We have described an ethically-justifiable, practical approach to decision-making surrounding separation of conjoined twins. In summary, decisions should be guided by a careful discussion of the family's goals of care, with careful attention to the benefits and burdens of associated with separation. Furthermore, the moral calculus used to separate conjoined twins must assess the interests of the twins from their perspective if possible, to avoid imposing the biases of a society unfamiliar with their experiences. We can find no moral justification to sacrifice the life of one twin to benefit the other.

Palliative care considerations

Palliative care, as defined by the World Health Organization, enhances quality of life through emotional, spiritual, social, and symptom support with interdisciplinary care teams in hospitals, clinics or in patient's homes.⁸ Palliative care improves the quality of life and symptom burden in patients with serious medical illness and should be offered in conjunction with cure-oriented, disease modifying care.^{8,9} Involvement of a palliative care team, if available, can be invaluable in the multidisciplinary care offered to conjoined twins. The palliative care team is an interdisciplinary group with the patient and family as its center. The team is comprised of physicians, advanced practice providers, nurses, social workers, chaplains, counselors, music and art therapists, child life specialists, pharmacists, volunteers, care managers and bereavement coordinators.

The tenants of palliative care, listed in Table 3, can be applied by any caretaker and are directly applicable to caring for conjoined twins. Palliative care can assist the medical team in developing goals of care and aid in complex medical decision making, beginning

prenatally and continuing throughout their medical care. Palliative care begins at diagnosis of illness. For conjoined twins, this is usually prenatally when fetal imaging discovers the condition. Overall prognosis for conjoined twins is poor, however the prognosis depends entirely on the anatomy and must be individualized to each set of twins.¹⁰ However, termination of pregnancy is frequently offered and often recommended to pregnant women with conjoined twins. This prompts a complicated decision for families that can be shaped by social, religious, political, and cultural influences. Palliative care can assist families in this decision by helping to clarify the family's goals for their twins, and helping to reconcile the factors that influence their decision. Risk for fetal death, potential maternal complications, concern for suffering and quality of life for the twins should all be taken into consideration. However, often there is uncertainty for potential of separation, which makes many of these factors such as quality of life difficult to truly comprehend.

For conjoined twins that progress to delivery, three possible scenarios exist including emergent surgical separation, delayed surgical separation, or no separation. Emergency separation often occurs when the anatomy of either twin threatens survival of one or both twins and when separation could potentially improve survivability for the healthier twin.^{11,12} When both twins are hemodynamically stable at birth and thereafter, the medical team must delineate anatomy to determine if separation is possible. After ensuring the family understands the risks and benefits of the surgery itself, along with outlining potential long-term outcomes, pursuing surgical separation is a complex decision that must take into account the family's goals of care. The palliative care team may provide anticipatory guidance to frame difficult and complex decision making related to surgical separation. In an effort to better delineate a family's goals of care, conversations may address each of the possible scenarios following surgery, including how to progress if one baby were to decompensate, for example. When a family chooses to pursue surgical separation, an elective delayed separation is preferred. However, either an elective or emergent surgical approach places one or both twins at risk for tremendous morbidity and mortality, likely with a prolonged hospital course and potential setbacks.^{10,13-15} An interdisciplinary palliative care team, when available, can offer additional support, psychosocial resources and continuity to patients and families. The palliative care team can also facilitate communication to ensure the parents' understanding and expectations are in line with the healthcare team's assessment and expectations.^{16,17}

Despite recent medical advancements, not all conjoined twins are candidates for surgical separation. The prognosis and ability to perform surgical separation on conjoined twins depend upon the prominent site of attachment.⁷ In these instances, the palliative care team affirms life while accepting death as a normal process, neither hastening nor postponing death. The palliative care team can help with memory making and legacy building, ensuring the family has a private, quiet area to spend time with their newborns, embracing and encouraging cultural or religious beliefs and traditions. If the conjoined twins are dying and surgical separation is not pursued, the palliative care team works to ensure that the medical interventions offered provide comfort and improve quality of life. This includes assisting in pain and symptom management, treating neuoroirritability, agitation, or agonal breathing, for example.^{17,18}

Conclusion

Conjoined twins pose unique challenges to ethical decision-making and to the provision of palliative care. Careful consideration must be given to the family's goals of care, the benefits and burdens of surgical separation, and the interests of the twins from their perspective as opposed to those of a society potentially biased against them. An interdisciplinary palliative care team, with the patient and family at its center, can help elucidate these considerations and navigate the ethical choices to arrive at a plan of care best suited for the twins and their family.

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REFERENCES

1. Wolfe J, Hinds PS, Sourkes BM. Textbook of interdisciplinary pediatric palliative care. Philadelphia: Elsevier; 2011.
2. Collins RT, Bhatti TR, Huff DS, Weinberg PM. Thoracopagus conjoined twins. *Circulation*. 2008;118:1496. [PubMed: 18824656]
3. London AJ. The Maltese conjoined twins: two views of their separation. *Hastings Cent Rep*. 2001;31(1):48–52. [PubMed: 11478094]
4. Wenkel DH. Separation of conjoined twins and the principle of double effect. *Christ Bioeth*. 2006;12:291–300. [PubMed: 17162676]
5. Kopelman LM. The best-interests standard as threshold, ideal, and standard of reasonableness. *J Med Philos*. 1997;22(3):271–289. [PubMed: 9232512]
6. Atkinson L. Ethics and conjoined twins. *Child Nerv Syst*. 2004;20:504–507.
7. Ross LF, Thistlethwaite JR. Minors as living solid-organ donors. *Pediatrics*. 2008;122(2):454–461. [PubMed: 18676567]
8. World Health Organization (WHO). WHO Definition of Palliative Care. [Website]. 2016 Accessed July 21, 2017 <http://www.who.int/cancer/palliative/definition/en/>
9. Kavalieratos D, Corbelli J, Zhang D, et al. Association between palliative care and patient and caregiver outcomes: a systematic review and meta-analysis. *JAMA*. 2016;316(20):2104–2114. [PubMed: 27893131]
10. Spitz L Conjoined twins. *Prenat Diagn*. 2005;25(9):814–819. [PubMed: 16170846]
11. Spitz L Ethics in the management of conjoined twins. *Semin Pediatr Surg*. 2015;24(5):263–264. [PubMed: 26382270]
12. Mian A, Gabra NI, Sharma T, et al. Conjoined twins: from conception to separation, a review. *Clin Anat*. 2017;30(3):385–396: (New York, N.Y.). [PubMed: 28195364]
13. O'Neill JA Jr., Holcomb GW 3rd, Schnauffer L, et al. Surgical experience with thirteen conjoined twins. *Ann Surg*. 1988;208(3):299–312. [PubMed: 3421755]
14. Spitz L, Kiely EM. Experience in the management of conjoined twins. *Br J Surg*. 2002;89(9):1188–1192. [PubMed: 12190687]
15. Brizot ML, Liao AW, Lopes LM, et al. Conjoined twins pregnancies: experience with 36 cases from a single center. *Prenat Diagn*. 2011;31(12):1120–1125. [PubMed: 21905053]
16. Munson D Withdrawal of mechanical ventilation in pediatric and neonatal intensive care units. *Pediatr Clin North Am*. 2007;54(5):773–785: xii. [PubMed: 17933622]
17. Kang TI, Munson D, Hwang J, Feudtner C. Integration of palliative care into the care of children with serious illness. *Pediatr Rev Am Acad Pediatr*. 2014;35(8):318–325: quiz 326.
18. Munson D, Leuthner SR. Palliative care for the family carrying a fetus with a life-limiting diagnosis. *Pediatr Clin North Am*. 2007;54(5):787–798: xii.

Table 1 –
Ethical permissibility of separation surgery based on pre- and post-surgical outcomes.

Pre-operatively: Both Die if Remain Conjoined, and Post-Operatively:						
Both Die		One Expected Survivor		Two Expected Survivors		
6 mos	> 6 mos	Major Morbidity	No Major Morbidity	Both with Major Morbidity	One with Major Morbidity	No Major Morbidity
Impermissible	Permissible	Maybe Permissible with Limitations	Obligatory with Limitations	Maybe Permissible	Permissible	Obligatory
Pre-Operatively Both Survive if Remain Conjoined, and Post-Operatively:						
Both Die		One Expected Survivor		Two Expected Survivors		
Impermissible		Impermissible		Both with Major Morbidity	One with Major Morbidity	No Major Morbidity
				Maybe Permissible	Maybe Permissible	Permissible to Obligatory

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Table 2 –

Criteria for principle of double effect.

1	The action in itself must be good or at least morally indifferent:
2	The agent must intend only the good effect and not the evil effect.
	The evil effect is foreseen, not intended
3	The evil effect cannot be a means to the good effect
4	There must be a proportionality between the good and evil effects of the action

Table 3 –

Tenets of palliative care, modified from the world health organization.

1	Affirm life while accepting death as a normal process
2	Intend to neither hasten nor postpone death
3	Offer a support system to help a families cope during a patient's illness and in their own bereavement
4	Interventions are aimed at comfort and quality of life
5	Consider values beyond the physical needs of a dying individual
6	Apply palliative care early in the course of illness in conjunction with other therapies intended to prolong life
7	Pediatric palliative care begins when illness is diagnosed and continues regardless of whether or not a child receives treatment directed at the disease

Adapted from World Health Organization definition of palliative care. Available at: www.who.int. Accessed June 5, 2007¹⁹