

## A Guide to Shared Decision-Making for Providers and Families

The challenges providers face in the prenatal care, delivery, and neonatal care of infants with Trisomy 13 and 18 translate into challenges in offering effective counseling for affected families. The complexities of the diagnosis of Trisomy 13 or 18 make understanding difficult especially if combined with life-threatening or life-limiting conditions for the fetus or infant. Families may be devastated or overwhelmed and often need time to absorb information and time to build trust with providers. Effective counseling can take multiple sessions with families over time. While extensive sessions with numerous members of the multi-disciplinary team may seem efficient, they often leave families without the time and reflection they need.

- Assessment of life-threatening anomalies must be taken into consideration for planning to be complete. A multi-disciplinary approach is key in assuring the family receives consistent messages. The message may change over time, as further information about the pregnancy or child becomes available, but the family must be able to trust that their providers' are approaching care together as a team.
- It is first important to gain an understanding of the family's goals for their infant (live birth, time at home with child, prolonging life, etc), including balancing factors such as the impact of treatments required to meet those goals (pain and suffering, hospitalization, medicalization, separation from family). The goal of the health care team is to understand how families perceive the *balance between burden and benefit* for their child in the context of medical or surgical options that in the providers' judgment may offer a benefit.
- Families may *change their minds* about what kinds of interventions are appropriate over time (both a change from minimal to more intervention and also from more intervention to less intervention) based on changes in outlook or understanding, new information about diagnoses, prolonged survival or other factors.
- It is helpful for some families to know about decisions they may face so that they can anticipate these challenges and think forward about what they might want for their baby as they gain experience caring for him/her.
- Ideally providers will present options to families based on the prospect of medical benefit and the family's preferences and goals in a *neutral, balanced and non-judgmental* manner. Support for the family's decision should be demonstrated (even if the option they chose differs from the recommendation or from what the provider would choose).
  - o Possible interventions may be framed as supportive, palliative, life-sustaining, or, perhaps more rarely, curative. The range includes 'comfort care', 'palliation', limited medical management, intensive medical management (invasive procedures) and/or surgical options

(after consultation with the surgical team). However, be very clear and specific about what is included in 'comfort care' or 'palliative care' (or avoid those terms altogether since these terms mean different things to different families and to different physicians).

- Questions to address *prenatally*:
  - a. What are options for further testing? Does the family wish to pursue those options? How would the family act on the information?
  - b. Does the family wish to continue the pregnancy? Note, counseling and decision-making may change as more information is gathered, as the pregnancy progresses and as the risk of fetal demise decreases.
  - c. As the pregnancy progresses, decisions about delivery planning are addressed. For families with a goal of seeing a liveborn infant, what kind of fetal monitoring will be done? Under what conditions will C/S or induction be offered? For C/S, convey the risks (maternal separation from infant, longer recovery and hospitalization, pain, long-term health consequences, etc) and benefits (potential for live birth in cases of fetal intolerance to labor, possibility of scheduling delivery date, etc.) in clear terms.
- Question to address for Delivery Room care: Does the family desire only comfort care (see Comfort Care Guide)? NCCC provider presence for confirmatory diagnosis based on physical exam findings followed by discussion of resuscitation measures? Limited resuscitation? Full resuscitation? CPR? What kinds of respiratory support? (CPAP, non-invasive ventilation, or a trail of invasive ventilation). Under what circumstances will NCCC admission be considered, recognizing that NCCC admission will separate the baby and family?
- Prematurity: discussions with families for infants where premature birth appears likely should cover the usual complications of prematurity. For example:
  - RDS: Understand the family's view of the risks and benefits of respiratory support such as nasal cannula oxygen, HFNC, CPAP, non-invasive ventilation, and intubation.
  - Apnea: Apnea of prematurity, obstructive apnea and central apnea related to CNS dysfunction can overlap. Ideally, the family and caregivers would come to a clear understanding of interventions and timing.
  - Nutrition: Understand the family's view of the risks and benefits of interventions such as IVF, HAL, gavage feeds.
- Invasive procedures/Surgical options: The risks and benefits of anesthesia and surgery need to be carefully explained to the family in close collaboration with the surgery and anesthesia teams. [For procedures currently considered at UNC, see 'Surgery/Invasive Procedures'].  
Issues and questions to address:
  - What are the goals of the procedure or surgery?

- a) Facilitate discharge? Full repair? Palliate (i.e. not complete repair but improvement in function or ease of care)? If palliative, what is the plan if there is long-term survival?
    - b) Learn more about a diagnosis? If so, how does what we learn help the family reach their goals?
      - Are there medical options for treatment that should be considered as an alternative to surgery?
      - What are the risks? What other anomalies or diagnoses are present that may complicate the surgery? For example, infants with Trisomy 13 or 18 may not extubate as quickly as infants without that underlying medical diagnosis and/or may require a longer hospitalization. Does that possibility affect the parent's view of risk and benefit?
  - Supportive Care Team: Works collaboratively with the medical team to assist with:
    - Understanding and clarifying the family's goals and wishes.
    - Helping families express worries and concerns about the medical condition itself or troublesome symptoms.
    - Supporting families in medical decision-making
    - Identifying resources in the home community.
  - Discharge feeding plan options:
    - Oral feedings, cup feeding, syringe feeding.
    - Home gavage feeds: Considerations –
      - What happens if the tube is dislodged at home?
      - Will the family learn go place the gavage tube? If yes, who will do the teaching?
      - Who will manage the feeding plan (increasing feeding volume)
      - Can the infant work on oral feeding?
      - If there is long-term survival, what is the next step?
    - Gastrostomy tube:
      - The family needs to be aware that infants with Trisomy 13 and 18 may not extubate as quickly as infants without this diagnosis.
      - Pain management needs to be clearly outlined.
- Offer appropriate hope and support. This may be as simple as offering hope for a livebirth or hope for a successful discharge, hope for a few days at home, etc., if these goals are reasonable.

- It is usually best to present options in a step-wise manner – for some families a few steps at a time, for others, projecting as far into the future as possible (given the clinical situation) is best. Parents who chose to proceed with treatment must address immediate life-threatening and life-limiting issues first. For some it is best to see how the child responds before deciding about other interventions. For example, making a decision about closure of a VSD is not a ‘required’ decision unless the child survives long enough for the VSD to become symptomatic. However, families may differ in the kinds of information they wish to hear and when. Whenever possible, it may be best to offer them a choice regarding the timing of such discussions.
- Assess the family’s support system: family members, community, church etc.
- Understand that families may seek advice and guidance from their own family and from other Trisomy 13/18 families in person or through social media. These patient narratives are powerful stories that we must account for in our conversations with families. The ways that families endorse (or criticize) the stories, or emphasize aspects of them, may help providers better understand the hopes or concerns that families may have. Offer families the chance to discuss the advice they have received from others (their family, extended family, colleagues, parents of children with Trisomy 13 & 18) as it may highlight parental concerns and the kinds of advice they value. It may help providers tailor their approach to meet the needs of the family.