

# Case Presentations in Perinatal and Neonatal Palliative Care

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# Case 1: Congenital Brain Malformation

I was asked by our NICU team to meet with a Mom who was diagnosed in her second trimester of pregnancy as having an infant with holoprosencephaly and bilateral cleft lip and palate without other apparent anomalies. Chromosomes obtained by amniocentesis were reported as normal XY male. She was referred to a Maternal/Fetal Medicine specialist for further care. At our initial meeting she and Dad expressed concern about what options would be available for both immediate post-delivery management as well as for home support if he survived the immediate neonatal period. They expressed the desire for their baby to not suffer unnecessarily and want to foster having as much quality time with him as possible. They felt they needed assistance to help them through this process, as well as to know what information to share with their two other healthy children – ages 3 and 6. Mom reported significant issues with anxiety and insomnia since being told of the infant's diagnosis.

# Case 1: Issues/Concerns

- **Impact on the family of this prenatal diagnosis**
- **Diagnosis of holoprosencephaly** – failure of division of the brain into right and left hemispheres early in gestation; there are multiple types with varying prognoses depending on the type
  - Alobar – worst type with most infants being stillborn, dying during delivery or soon after birth, rarely surviving 6-12 months of life
  - Semilobar/Lobar – intermediate types with 50% of children without significant malformations of other organs alive at age 12 months
- **Planning for Perinatal/Neonatal Management**
- **Care after the hospital if infant survived the immediate newborn period**

# Case 1: Progression

- Parents felt they needed clarification of the diagnosis and expected prognosis to assist them in decision-making
  - Role of the Internet and Information Accessibility
  - Advanced testing – some available only after the infant’s delivery
    - High-level Ultrasound
    - Amniocentesis with chromosome analysis
    - Fetal MRI
    - Microarray chromosomes
- Options for Delivery Room/NICU management needed to be discussed, emphasizing avoiding “crisis mode” decision-making while allowing for them to “change their mind” at any point
- *Introduction* to expected medical complications “down the road” and options for management and support

# Case 1: Progression

- Parents, given ambiguous information with the full extent of prenatal testing available, elected for a Birth Plan specifying aggressive NICU care including intubation/mechanical ventilation if needed – to be followed by further diagnostic testing in the NICU to help determine further goals of care and treatment.
- Infant did well in L&D, requiring only O2 via N/C. MRI of his head revealed semilobar holoprosencephaly. OG/NG feedings were initiated and tolerated fairly well. Another family meeting discussed transition to home and availability of hospice support. Family elected to go home with a cardio-respiratory monitor, hospice care, with an ***Allow Natural Death*** order in place.

# Case 1: Progression

- He expired at home during his 3<sup>rd</sup> week of life after a brief period of respiratory distress which was managed with comfort medications, O2 along with family support from his hospice nurse. Mom expressed joy at having been given the opportunity to bond with him, albeit a short time.
- Results of microarray chromosome analysis (from blood drawn while in NICU) showed a partial deletion of Chromosome 18. Mom was referred to Genetics for further evaluation and counseling, including assessment of recurrence risk.
- Family received bereavement support services from the hospice team which included notifying the older child's guidance counselor and teacher regarding the sibling's death.

## Case 2: Multiple Congenital Anomalies

- I was contacted by the NICU team to meet with the family of a 5mo infant who was struggling emotionally because their baby had developed progressive respiratory failure and episodes of severe agitation associated with severe oxygen desaturations (and now bradycardia) which were only minimally responsive to treatment with opioids and benzodiazepines. He was now being treated with neuromuscular blockade (paralysis) in order to provide adequate ventilation. He was born with an omphalocele and had resultant severe pulmonary hypoplasia. He has now developed pulmonary hypertension which has proven unresponsive to usual treatments (sedation, sildanefil, nitrous oxide). He needs an operative procedure for repair of his primary abdominal wall defect but may not survive to have that surgery.

## Case 2: Issues/Concerns

- **Declining clinical course** – the NICU “roller-coaster”
- **Quality of life** – pulmonary requirement for neuromuscular blockade, difficulty of assessing for adequate sedation in a paralyzed infant, NICU caregiver “angst”, parents wanting to be able to “interact” with their infant even if he would only tolerate it for brief intervals
- **Hope for eventual repair of congenital defect and “normal” life**
- **Differences in parental “coping” with declining course and therefore difficulty in coming together during *goals of care* discussions**



## Case 2: Progression

- Multiple options for sedation were tried with some success including methadone with slow taper, phenobarbital, scheduled and prn benzodiazepine regimens
- Family established *Allow Natural Death* status and rescinded that status for *Full Code* multiple times during his prolonged NICU hospitalization
- NICU Plan of Care was modified to allow for his paralysis to be stopped during parental visits to allow meaningful interaction between the parents and their infant
- Parents, NICU team, specialty physicians and palliative care had multiple meetings to ensure family was kept informed regarding his progress (or lack thereof) and emphasized the desire to ***honor the care they wanted for their child.***

## Case 2: Progression

- Ultimately, he “outgrew” his lungs and progressed to death despite all efforts. Parents remained very attentive throughout his NICU stay and were able to introduce him to his older brother (“bending” NICU rules) and enjoy meaningful interactions with him.
- Parents expressed feeling they had “failed” to protect their child from death but responded to comfort from the NICU team and palliative care pointing out the value of their continued consistent presence
- Role for a “Parent Medical Record” to track progress and help parents “see”/accept trajectories of disease

## Case 3: Uncertain Prognosis

- I was asked to see this 5 week old infant - after an extensive NICU evaluation for multiple congenital anomalies – to provide palliative care follow-up and assist with “transition of care” planning. Born after a “normal” pregnancy, she was noted to have dysmorphic features. Her congenital anomalies included partial agenesis of the corpus callosum, congenital scoliosis, hypoplastic optic nerves, multiple joint contractures, airway anomalies causing chronic stridor and intermittent respiratory distress, and chromosome analysis showing an unbalanced translocation of the 15<sup>th</sup> chromosome. She was being treated for hypertonicity with PT and Baclofen, was requiring NG feedings for severe swallowing dysfunction per MBSS. She was going to need follow-up care with multiple specialists including Neurology, Orthopedics, Ophthalmology, ENT, as well as routine Pediatric outpatient care. Mom was already frustrated at communication issues with specialty physicians and was worried about being able to adequately care for her at home.

## Case 3: Issues/Concerns

- Rare chromosomal abnormality with resultant multiple congenital abnormalities but with **unknown long-term prognosis** though all felt she would have a limited life expectancy and certainly significant morbidity
- **Parental discomfort with required level of care**
  - Psychosocial issues
  - Other siblings in the home
  - Learning NG tube placement, feeding skills
- **Need for Coordination of Care** both in the hospital and outpatient setting to include physician and support services such as PT, OT
- **Maternal concern about medication side effects**

# Case 3: Progression

- We discussed the difficulty of prognostication considering each of her multiple congenital anomalies as well as the “overall picture”
  - Utilize prognoses of the individual anomalies to help
    - Seizures, neurodevelopmental delay with agenesis of the corpus callosum
    - Likely inability to walk due to difficulty of ameliorating the congenital joint contractures
    - Severe visual impairment but possibility of improvement over time
  - Describe difficulty of one problem exacerbating another
    - Congenital scoliosis may impair respiratory function as well as chronic aspiration from swallowing dysfunction
    - Airway anomalies and swallowing dysfunction would affect feeding issues and respiratory function
- Encouraged a “Wait and See Approach”
- Discharge Planning

## Case 3: Progression

- Discharge planning was done including Mom “rooming in” to ensure comfort with medication administration. A G-tube was placed prior to NICU discharge due to Mom’s discomfort with NG feedings.
- Phenobarbital and Baclofen were discontinued with improvement in her level of alertness which pleased Mom. PT/OT was continued to decrease risk of further contractures.
- Referrals for Early Intervention, SSI and Katie-Beckett Medicaid were completed
- Palliative Care provider facilitated communication between her PCP and the multiple specialty physicians via direct communication, secure email “discussions” and transmission of summary notes from the EMR to various providers

## Case 3: Progression

- She is now 10 months old. She has continued to have airway issues and lays with her neck chronically hyperextended. An airway evaluation revealed very poor handling of oral secretions with essentially funneling of oral secretions toward her airway – despite this, she has not had any episodes of clinical aspiration pneumonia. She is able to roll side to side, tracks well visually, and is clearly interactive with her caregivers. She had a markedly abnormal sleep study but Mom elected not to proceed with tracheostomy despite understanding the risks because she felt her daughter would be more comfortable without this. A brace was prepared to try to prevent worsening of her congenital scoliosis but she did not tolerate it at all due to respiratory compromise so Mom is not using this for now. Her scoliosis has con't to worsen. Palliative care has con't to provide support for Mom and baby and assists with coordination of care.

## Case 4: Unrealistic Expectations

- Born at 24 weeks gestation after several weeks of bedrest for incompetent cervix to parents who had several attempts at IVF before conceiving her, she had a birth weight of 490 grams. She had a complicated first week in the NICU with need for significant ventilatory support, skin breakdown, electrolyte imbalances, hypotension and the development of bilateral Grade IV intraventricular hemorrhages. Parents have been resistant to reorienting the goals of care toward palliation despite her dismal prognosis and multiple meetings with NICU and support staff. Palliative care is consulted and is asked to address some specific questions/concerns as follows.



## Case 4: Issues/Concerns

- How can the NICU team help the parents gain and accept understanding of her declining course and its implications for her long-term prognosis?
- What supports can be put into place to help these parents as they come to terms with the inevitable outcome for their daughter?
- If the parents choose to withdraw artificial support, how can the NICU and Palliative Care team make that as peaceful and meaningful a time as possible?
- After the baby dies, what can be done to help the family?

## Case 4: Progression

- Review of “hard findings” with parents (looking at serial head ultrasounds or CTs, reviewing chest x-rays) as well as reiterating formal assessments by specialty physicians
- Developing a parent “logbook” to help demonstrate decline despite maximal support
- Encourage parents to seek their support network (family, faith) to help them in this time of stress
- Acknowledge their grief (anticipatory) at the potential loss of this long and much-desired infant

## Case 4: Progression

- Offer “Memory Making” assistance including professional photographer, cutting locks of hair to save, allowing them to “parent” as much as possible – holding their infant and caring for them
- Provide a more private environment for them to spend the limited time they have with their infant, particularly when withdrawal of artificial support is planned
- Ensure parents understand that comfort medications will be given to ease transition off of artificial support
- Allow family to bathe, dress and prepare their infant’s body for funeral home delivery
- Ensure long-term support via hospice bereavement, follow-up calls from NICU staff

# Summary of Important Principles

- Talk honestly with families regarding diagnoses and prognosis (acknowledging uncertainty if present) and assess their understanding
- Emphasize your desire to “honor their wishes for their baby”
- Encourage delineation of goals of care and ensure the plan of care is congruent with the goals
- Emphasize “quality” of life over “quantity” of life, being careful to not impose your views of “quality” on the family
- Integrate the baby into the family by avoiding “technology isolation”
- Communicate in an ongoing and caring fashion even when parental decision-making doesn’t “match” that of the medical team