The Care of the Medically Complex Child &Their Transition to Adult Medical Care

Diana Kudes, MD
Jessica Mayer, DO
POFPS 43rd Annual CME Symposium
August 3, 2018

Disclosures:

Dr. Kudes has provided no disclosures.
Dr. Mayer has provided no disclosures.

#POFPS43

Goals

Help family physicians be more comfortable in the care of the medically complex pediatric patient.

Help family physicians transition care of the medically complex child to adult care.

Help family physicians assume care of medically complex children transitioning from pediatric care.

#POFPS43

Essential Tools and Supports

- Good communication with other medical providers and specialists
- Knowledge of the common problems associated with the disorder
- Information about community resources
- Awareness of the parent's own emotional and medical needs

#POFPS43

Complex Pediatric Patients Examples

Prematurity

Down Syndrome

Autism Spectrum Disorder

#POFPS43

The Premature Infant
(Born before 37 weeks)

Seen 24-48hrs after discharge from the hospital

- Hospital discharge records are essential to review at the visit to determine follow up visits that are needed, medications, medical equipment, immunization records, and growth.
- Carefully review feeding history including type and amount of feeds.
- If weight gain is adequate another visit should be scheduled in 2-4 weeks.
- Subsequent well child visits based on the child's chronological age.

#POFPS43

The Premature Infant
Follow Up Specialists

- Ophthalmologist
- Pulmonologist
- Cardiologist

#POFPS43

The Premature Infant

- Immunizations are given based on the baby's chronological age.
- Growth and development are based on the child's corrected gestational age(CGA). So they should be plotted based on CGA until age 2yrs.
- WHO growth curves are appropriate for low birthweight infants (LBW) or babies less than 2500g.
- Very low birthweight (VLBW) or babies less than 1500g may be plotted on WHO growth charts or IHDP growth charts for premature infants. There are some criticisms of both charts for this group.

#POFPS43

Breast Feeding

- Breastmilk is the best nutrition for the premature infant however breastfed infants tend to gain weight slower than formula fed infants.
- In the NICU babies are usually given breastmilk with fortifiers to increase weight gain.
- Evidence for continuing fortification after discharge are small but are recommended by many NICUs. Fortifiers are continued until corrected weight is 25% or greater or 6 months.
- The European Society for Paediatric Gastroenterology, Hepatology, and Nutrition recommends fortification for exclusively breastfed infants with subnormal weight for corrected gestation age. [2]

#POFPS43

Feeding

- Formula fed infants are fed enriched 22cal/oz formula in the NICU for improved growth and nutritional needs.
- There is no data to support continuation of enriched formula for all former premature infants.
- Infants who are <10% at NICU discharge are usually discharged on an enriched formula until they are >25% or 6 months old.
- Infants should usually be allowed to eat on demand every 2-4hrs for a total of 120-150ml/kg/d.

#POFPS43

The Premature Infant: Palivizumab

- Depending on how premature the infant is and other comorbidities the infant may qualify for palivizumab. The AAP has published guidelines for its use and reviews its policy yearly.
- Infants in the first year of life who were born before 29 weeks qualify but infants born >29 wks may qualify if certain conditions are met.
- RSV Policy Statement —Updated Guidance for Palivizumab Prophylaxis Among Infants and Young Children at Increased Risk of Hospitalization for Respiratory Syncytial Virus Infection. *Pediatrics* 2014;134(2):415–420 contains the current guidelines

#POFPS43

The Premature Infant: Iron Deficiency

- Preterm infants have low iron stores, decreased erythropoietin production, shortened RBC survival and have often had frequent venipuncture putting them at higher risk of developing anemia.
- Breastfed preterm infants should be given 2mg/kg of iron from age 1mo to 12mo or until getting adequate iron intake from foods
- Screen for iron deficiency anemia in all preterm infants between ages 4-8 months, and then again at 12 and 24 months of age.

#POFPS43

The Premature Infant: Development

Development should be based on the corrected gestation age and evaluated at every well child check. Any delays should prompt referral to Early Intervention.

Referral to Early Intervention surveillance programs may be initiated at NICU discharge. Most NICUs also have follow up clinics which will include development surveillance.

#POFPS43

Premature Infant:

Premature infants are at higher risk of

- Cerebral palsy
- Motor and coordination problems
- Cognitive impairment
- ADHD

#POFPS43

Down Syndrome

The AAP published guidelines for the health supervision of the child with Down Syndrome from birth to age 21 years in 2011.

These guidelines include useful information about additional health screenings and anticipatory guidance unique to this population of patients.

#POFPS43

Down Syndrome: Initial newborn visit

Review of the hospital discharge is critical and should include:

- How the diagnosis was made
- Evaluation for congenital heart disease including echocardiogram
- Need for feeding evaluation
- Need for urgent Ophthamoligic follow up
- Hearing evaluation
- Results of CBC
- Newborn screen results especially thyroid
- Do they have a condition that qualifies them for RSV prophylaxis

#POFPS43

Down Syndrome: Growth & Development

Infants and children should be monitored closely for growth and development.

World Health Organization Growth charts should be used instead of Down Syndrome charts previously recommended because they no longer reflect current population growth.

Children with Down Syndrome are typically of shorter stature and heavier than other children.

Intellectual disability can be mild to severe.

#POFPS43

Down Syndrome: Common medical problems

- Obstructive sleep apnea
- Ear infections and hearing loss
- Cataracts and visual problems
- Congenital heart disease
- Thyroid disease
- Seizures
- Hip disorders
- Congenital Gastrointestinal malformations
- Autism
- Celiac disease
- Hematologics problems: Myeloproliferative disorder, Leukemia
- Atlanto-Axial instability

#POFPS43

Routine Screenings and Referrals After Newborn Nursery

- Refer to Pediatric Ophthalmologist in the first 6 months. Annual visits ages 1-5 yrs, then every other till age 13yrs, then every 3yrs.
- Repeat Thyroid studies at 6mo, 12 months and then annually
- Refer to Audiology at 6 months
- Hearing testing and tympanometry every 6 months from ages 1-5yrs, then hearing testing annually ages 5-21yrs.
- Hgb annually starting at age 1yr.
- Refer for sleep study by age 4ys.

#POFPS43

Autism Spectrum Disorder

- The American Academy of Pediatrics recommends that every child should be screened for autism at 18 months and 24 mo using the Modified Checklist for autism in toddlers revised version or M-CHAT-R/F.
- The American Academy of Family Physicians has stated there is insufficient evidence to screen everyone in lines with the recommendations from the US Preventative Task Force.
- The most recent data from the CDC from 2014 show that 1 in 59 children have autism.

#POFPS43

Autism Spectrum Disorder: Comorbidities

- ASD is more common in children with Down Syndrome, Fragile X syndrome and Tuberous sclerosis.
- ASD occurs with another developmental diagnosis 83% of the time.
- Psychiatric diagnosis is also present in 10% of children.
- Vanderbilt University has a toolkit for Health Care For Adults with Intellectual and Developmental Disabilities.
- About half have an intellectual disability

#POFPS43

What to Watch For: HEENT

- Otitis media
- Allergies
- Hearing loss
- Sensitivity to sound and light
- Cavities

#POFPS43

Autism Spectrum Disorder: Sleep

- Sleep problems are very common.
- Obtain detailed history
- Night waking may be from OSA, restless legs, seizures, sleep walking
- Sleep hygiene can be reviewed
- Vanderbilt has handouts for families on sleep tips that are useful.
- Consider referral for sleep study

#POFPS43

Autism Spectrum Disorder: Gastrointestinal problems

- Constipation
- Diarrhea
- GERD
- Food preferences
- Food allergies
- Ask about special diets or alternative treatments they may be trying

#POFPS43

Autism Spectrum Disorder: Neuro/Muscular

- Hypotonia
- Seizures
- Gross Motor delay
- Fine Motor delay
- Motor planning problems
- Tic disorders

#POFPS43

Autism Spectrum Disorder: Behavior and Emotion

- Poor social skills
- Wandering
- Sensory issues
- Tantrums and self injurious behaviors
- Depression
- Anxiety
- OCD
- ADHD

#POFPS43

Infectious Disease

- Otitis media
- 25% have impaired immunity

#POFPS43

Autism Speaks

- Resources for parents and providers
- Resources for children and adults with ASD
- Support staff in English and Spanish with global links as well.
- Has links to find services in your local community
- Connects parents with other parents
- Accurate information

#POFPS43

Transition to Adult Medical Care

- For conditions such as Down Syndrome and Autism special care should be taken to ensure a smooth transition
- May need to visit the office prior to having their first appointment to be comfortable with a new environment
- Providing a clear explanation of what to expect during an appointment can help reduce anxiety.
- This can be verbal, written or through pictures or a combination. Even patients who are literate may benefit from pictures.

#POFPS43

Transition to Adult Medical Care

- During appointments, be sure to explain everything that is about to be done to the patient, even things we may think are routine or have been done to the patient before
- Be sure to address the patient directly, do not only speak to caregivers
- Often helpful to schedule longer appointments
- Important to establish baseline as minor changes may signify significant medical problems
- Ability to communication symptoms or pain may be limited

#POFPS43

Medical Conditions with higher prevalence in Adults with Down Syndrome

- In addition to those previously referenced, in adulthood the following medical conditions are more prevalent:
  - Mental Health Disorders: Depression, Obsessive Compulsive Disorder, Conduct Disorder
  - Abuse (physical and sexual)
  - Alzheimer's disease
  - Testicular cancer
  - Diabetes
- Medical problems tend to manifest as behavioral problems
- All standard adult screening should be done

#POFPS43

Special Considerations

- An adult with Autism or Down Syndrome is considered competent to make their own medical decisions unless specifically declared otherwise
- In cases where guardianship is not appropriate, it is important to address advance directives, especially power of attorney for health care and finance
- Family estate planning is important, frequently a trust needs to be established

#POFPS43

Resources

Autism Services in Pennsylvania
http://www.dhs.pa.gov/citizens/autismservices

Autism Speaks http://www.autismspeaks.org/

Autism Case Training. Centers for Disease Control and Prevention. https://www.cdc.gov/ncbddd/actearly/autism/case-modules/index.html

General Guide for treating Adults with Disabilities
Adults with Developmental Disabilities: A Comprehensive Approach to Medical Care. *American Family Physician.* 2018;97(10):649-656

#POFPS43

References

1. "Autism Spectrum Disorder: Data and Statistics." Centers for Disease Control and Prevention, April 26, 2018.
2. "Final Recommendation Statement: Autism Spectrum Disorder in Young Children: Screening." US Preventative Services Task Force, September 2017.
3. "Health Care For Adults With Intellectual And Developmental Disabilities: Toolkit for Primary Care Providers: Health Watch Table-Autism". Vanderbilt Kennedy Center. 2018 (https://vkc.mc.vanderbilt.edu/etoolkit/physical-health/health-watch-tables-2/)
4. Supporting the Health Care Transition From Adolescence to Adulthood in the Medical Home. American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians, Transitions Clinical Report Authorizing Group *Pediatrics* 2011;128;182
5. Bull, M and the Committee on Genetics. Clinical Report-Health Supervision for Children With Down Syndrome. *Pediatrics*. 2011;128;393-406.
6. Bunt C, Bunt S. Role of the Family Physican in the Care of Children with Down Syndrome. *American Family Physican* 2014;90(12):851-57
7. Gauer R, Burket J, Horowitz E. Common Questions About Outpatient Care of Premature Infants. *American Family Physican Physician*. 2014;90(4):244-251.
8. www.cdc.gov/growthcharts/growthchart_faq.htm
9. Platt, MJ. Outcomes in preterm infants. *Public Health*. 2014
10. Sherry B, Mei Z et al. Evaluation of and Recommendations for Growth References for Very Low Birthweight (<1500g) Infants in the United States. *Pediatrics* 2003;111;750
11. Autism Spectrum Disorder: Primary Care Principles. *American Family Physician*. 2016; 94(12):972-979
12. Health Care Management of Adults with Down Syndrome *American Family Physician*. 2001; 64:1031-8, 1039-40

1. "Autism Spectrum Disorder: Data and Statistics." Centers for Disease Control and Prevention, April 26, 2018.
2. "Final Recommendation Statement: Autism Spectrum Disorder in Young Children: Screening." US Preventative Services Task Force, September 2017.
3. "Health Care For Adults With Intellectual And Developmental Disabilities: Toolkit for Primary Care Providers: Health Watch Table-Autism". Vanderbilt Kennedy Center. 2018 (https://vkc.mc.vanderbilt.edu/etoolkit/physical-health/health-watch-tables-2/)
4. Supporting the Health Care Transition From Adolescence to Adulthood in the Medical Home. American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians, Transitions Clinical Report Authorizing Group *Pediatrics* 2011;128;182
5. Bull, M and the Committee on Genetics. Clinical Report-Health Supervision for Children With Down Syndrome. *Pediatrics*. 2011;128;393-406.
6. Bunt C, Bunt S. Role of the Family Physican in the Care of Children with Down Syndrome. *American Family Physican* 2014;90(12):851-57
7. Gauer R, Burket J, Horowitz E. Common Questions About Outpatient Care of Premature Infants. *American Family Physican Physician*. 2014;90(4):244-251.
8. www.cdc.gov/growthcharts/growthchart_faq.htm
9. Platt, MJ. Outcomes in preterm infants. *Public Health*. 2014
10. Sherry B, Mei Z et al. Evaluation of and Recommendations for Growth References for Very Low Birthweight (<1500g) Infants in the United States. *Pediatrics* 2003;111;750
11. Autism Spectrum Disorder: Primary Care Principles. *American Family Physician*. 2016; 94(12):972-979
12. Health Care Management of Adults with Down Syndrome *American Family Physician*. 2001; 64:1031-8, 1039-40

#POFPS43