



# **TRANSITION CARE**

## **(A paediatric aspect)**

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# History

- MM-16 yrs 5mths
- Normal birth and normal development
- Regression of milestones at ~4 yrs
- Not investigated- maternal refusal
- Admitted with seizures at 8 years
- Investigated for Developmental delay



# Diagnosis

- Sanfilippo A Syndrome
- Mod-severe intellectual delay
- Language regression
- Behavioural issues/Aggression
- Epilepsy
- Sleep disturbance
- Psycho-social issues, Families SA



# Sanfilippo Syndrome

- Described in 1963 by Dr. S. Sanfilippo
- Lysosomal Storage Disorder
- MPS 3 (4 main types A-D) A- severe
- 1 in 70,000 births
- Neurodegenerative disorder
- CVS for prenatal diagnosis
- Treatment-SUPPORTIVE



# Disease Profile

- ~3years- Initial normal development
- Delay in milestones “slip backwards”
- 5-10 years -Challenging behaviours
- >10 yrs - slow, ataxia, wheelchair bound
- 14yrs -bedridden, ‘dementia’
- Death teens to early twenties.



# Other Clinical Issues

- Conductive deafness
- Increased respiratory infections
- Bouts of severe diarrhoea
- Constipation
- Spasticity
- Swallowing difficulties
- Anaesthetic problems
- Cardiomyopathy



# Current status

- Some eye contact
- Claps hands
- No words/no drooling
- Ataxia-upper limbs, unsteady gait
- Wheelchair for mobility
- Ongoing intermittent seizures
- Attends special school
- Psychosocial issues/ refusal to treat



# Transition Issues

- Definition: (Am Soc for Adol Med)
  - Purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centred to adult-oriented health care

## Why?

- Increasing survival rates of complex diseases
- Advance in medical technology



- **Childhood**

- Coordinated and multidisciplinary
- Paediatrician

- **Adulthood**

- ? Risk of inadequate/fragmented healthcare
- No readily accessible adult equivalent to paediatrician
- GPs may be overwhelmed
- Multiple referrals to specialist and hospital visits



# Barriers

- Emotional, long standing relationship with paediatrician
- Paediatricians view as “experts”
- Paediatricians “underestimate” adult caregivers knowledge and skills
- Differences in practice styles and philosophy



# Goals

- High quality
- Uninterrupted
- Coordinated healthcare
- Patient centred
- Age and developmentally appropriate
- Comprehensive
- Flexible




# Transition issues for MM

## Behind the scene

- ‘prime the family’ - “letting go”
- Identify unit/doctor in adult hospital
- Commence communication lines
  - Paediatrician to Physician
  - Paediatrician to Parent
  - Parent to Physician

Multidisciplinary case conference

“Everyone should be on the same page”

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- Respect the choices made by the family
  - Case summary as well as general information about the specific condition
  - Special Needs folder in ED of adult hosp
  - Issues of consent (family/GOM)
  - Issues regarding current resuscitation orders-  
clear documentation



# Hospitalisation-Key Issues

- Acute/uncontrolled seizures
- Behavioural issues/dementia
- Respiratory infections
- Feeding difficulties and Inanition



# Hospital Issues

## Apart from medical management

- Nursing needs 1:1 nursing
- Palliative Team introduction
- Physiotherapy/Occupational Therapy
- Social Worker
- Dietician
- Ongoing input-Novita



# Epilepsy

- Treatment similar to anyone with seizures
- Further intervention as per resus plan
- Intubation-use agents that minimise post-extubation behavioural problems
- 1:1 nursing (violence)

Re-address role of maintenance anticonvulsant therapy



# Screaming episodes

## To exclude medical causes

- Otitis media (very common)
- Constipation
- Urinary tract infection
- Dental abscess
- Dislocation of hips
- ? Fractures ??NAI



# Respiratory infections

- Clearly defined treatment plan
- Increasing oxygen requirement/deterioration
- Consent issues regarding ventilation
- Revisit dialogue re: palliative care



# Feeding difficulties

- Neuro-degeneration, with loss of gag and swallow reflex
- Frequent aspirations
- Alternative feeding (N/G or Gastrostomy)
- Dietician input
- Consent for procedure



## References

- Kliegman R, Muenzer J. Chapter 77 on Mucopolysaccharidoses in Nelson Textbook of Paediatrics 17th Ed. Saunders 2005
- Meyer A et al. Scoring Evaluation of the Natural Course of Sanfilippo Syndrome A Paediatrics 2007; 120:5:1255-1261

# References

- Constance TRM et al. Healthcare transition in persons with intellectual disabilities: General Issues, the Maastricht Model, and Prader-Willi Syndrome.  
Am J Med Genet 2007; 145C: 241-247
- Viner R. Transition from paediatric to adult care. Bridging the gaps or passing the buck?  
Arch Dis Child 1999; 81: 271-275



## References

- Transition to Adult health care for Adolescents and Young Adults with Chronic Conditions-Position Paper  
J of Adol Health 2003;33:309-311