

Intellectual Disability & Epilepsy across the Lifespan

Dr Helen Somerville

Children's Hospital Westmead

and

Westmead Hospital



Intellectual Disability

Cognitive impairment manifest before the age of 18 years.

UK = Learning Disability

US = Mental Retardation



Association of Epilepsy and I.D.

- Overall incidence 22% (Welsh study 1996)
- Cerebral Palsy 50%. Incidence increases with increasing severity
- Tuberous Sclerosis 100%
- Autism 30%
- Down Syndrome 30% (increase with age)
- Angelman 50%
- Fragile X 25%

Different Issues for the I.D. Population

- Diagnosis more difficult
- Impact on QOL different
 - Driving
 - Employment
 - Pregnancy
- Goals/expectations of treatment less clear
- Frequent co-medication

Co-morbidities

- G.I.T.
- Chest
- Mental Health/Behavioural Issues
- Dystonia/other neurological
- Vision/Hearing
- Endocrine –bone health + menstrual management
- Sleep disorders



What is transition?

“The purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centred to an adult oriented health care system”

Blum et al. (1993) J Adolescent Health

Transition (cont)

- Design not Default
- Planning/Preparation
- Patient stability
- Comprehensive medical history
- Summary for ED
- Contact details - Health Professionals
- Case Manager – eg GP
- Psychosocial issues

Transition (Cont)

- Full seizure, medication and Ix history
 - incl failed medications and adverse effects
- Parent/carer expectations
- End of life – Palliative care, not withdrawal of care



Health care challenges for people with ID

- Complex medical and psychosocial
- Communication
- Co-operation-may need GA for exam/Ix
- Consent
- Lack of continuity of care

Paediatric

- Paediatrician case mgr
- Variety of clinicians with ID expertise
- Consent by parent
- Multidisciplinary care
- Lifelong records
- Optimism
- Long association with clinicians & system
- Child and family centred

Adult

- No case mgr
- Little exposure to ID
- Consent by ?
- Subspecialist care
- Records often limited
- Realism
- Trust takes time
- Seen as less “warm”

What good transition hopes to avoid

- 1st contact via ED
- Poor transfer of medical records
- Communication difficulties
- Consent issues
- “Bad press” - perceived inability of adult service to “cope”

Cases from a Complex Epilepsy/Disability Clinic

45 Patients

– Cerebral Palsy	15
– Congenital/Childhood Infection	3
– Autism	3
– Chromosomal	4
– Rett	3
– SMEI	3
– Other genetic	8
– Tumour	2
– Unknown	2

Cases - Co-morbidities

• GORD	22
• Enteral Feeding	15
• Osteopaenia/osteoporosis	23
• Mental Health	16
• Other neurological e.g. dystonia	7
• Vision/hearing	10

Cases - Outcomes

- Re-evaluation of seizure diagnosis
- AED review
- Concomitant seizure-provoking meds
- Behaviour
- Re-evaluation of “unifying” diagnosis
- “Quality of Life” focus
- Change from “cure” to “management”
- Prognosis

Eliza – dob 18/4/89

- Spastic quadriparesis – severe ID
- Neurodegenerative - familial
- Epilepsy
- GORD – gastrostomy
- Chronic lung disease
- Osteoporosis
- Constipation
- Thalassaemia trait



Eliza – 1st presentation to adult hospital

- Prolonged seizure
- Respiratory compromise
- Probable aspiration
- Initial treatment order – NFR

Mickey – dob 24/3/82

- Down Syndrome
 - Epilepsy
 - Hypothyroidism
- Choking episode during sz
- Sudden deterioration in mobility/hand function
- Epilepsy management – acute and long term

Karrie – age 20 years

- Intractable epilepsy – 3 seizure types
- Double cortex syndrome
- Menstrual issues
- Multiple anticonvulsant trials – side effects ++ or are they?... VNS being considered
- “Teenage” behavioural issues
- Maternal mental health

Practice Points – Epilepsy & ID

- History worth the extra effort!
- Same rules of management for all
- Specify what info. you require
- Define recording & monitoring incl sz charts
- Specifics – bathing/swimming
- YOUR “Epilepsy Management Plan”
- Communication with colleagues, esp GP
- Role of nurse specialist

Group Home Issues

- Kylie and Bruce
- Level of expertise
- Little or no relevant training
 - Role of specialist nurses
- Paperwork/rules
- Lack of corporate memory
- Lack of continuity of care
- Doctor shopping
- ☺ Many excellent carers & group homes!

Glitches in Practice

- Urgent transition
- Consent issues
- No medical records
- No case manager
- GA/pain relief needed for examination/Ix
- Turf battles/Funding issues
- Lack of mental health services
- NFR orders by adult ED teams

Traps

- Attributing behavioural issues to
 - Epilepsy
 - Medication changes
- Pseudoseizures DO occur in the ID population
- Treating the EEG – 90% abnormal
- Treating the seizure record
- Effect of antipsychotics

