

Palliative treatment in the NCLS

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Fulda, January 2006

Summary

- Introduction
- Principles of care
- Juvenile onset NCL
- Late infantile onset NCL

History

- Antioxidants (Santavuori, et al 1988)
 - Vitamins E & C, methionine, selenium
 - no benefit on vision
 - no definite effect neurological/cognitive decline
 - Mnd mouse model – no benefit with Vit E supplements (Griffin et al, 2002)
- PUFA (Bennett et al 1997)
- Carnitine (Katz et al 1997)
- Flupirtine

Principles of care

- No harm
- Optimise quality of life
- Holistic

- Manage symptoms
- Maintain and promote skills

- Partnership with individual
- Partnership with family and carers
- Partnership with multidisciplinary colleagues

Considerations

- Environment
- Physical and emotional needs
- Medical needs
- Day to day variability
- Changing needs
- Respite for carers

Juvenile Batten Disease (NCL)

- Vision
- Seizures
- Mobility and spatial awareness
- Communication
- Emotional health
- Mental health
- Bladder and bowels

Seizures

- Seizures all kinds
 - Commonly GTCS
 - Subtle partial, episodes of partial status unrecognised
 - ?relationship with hallucinations/variability in skills
- Valproate, Lamotrigine, Topiramate, Clobazam, Leviteracetam
- Aim for good control
- Drug interactions with antipsychotics
- Role of rectal/buccal benzodiazepines or paraldehyde

Mobility and spatial awareness

- Characteristic posture
- Slow Parkinsonian gait, difficulty sitting back in chair, initiating movement
- Easily lose balance, may fall
- +/-Tremor
- Effects on speech – difficulty initiating speech
- Variability from day to day

Mobility and spatial awareness

- Seating and posture
- Lifting and handling
- Enjoyable and therapeutic activities
 - enhance well-being, maintain posture and skills, prevent injury and deformities
- What is the place of Anti Parkinsonian drugs?

Anti-Parkinsonian drugs

- Sinemet/Madopar
 - L-Dopa combined with extra cerebral dopa-decarboxylase
 - Long term extra pyramidal effects
 - Drug interactions with antipsychotics
- Heather House experience:
 - used early to enhance spatial awareness and feelings of safety and wellbeing
- Aberg et al 2001

Communication

- Repetitive, staccato speech, often pressured
 - Becomes more difficult to understand
 - Recurrent conversation themes
 - Frustration
 - Variability from day to day
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- Importance of memories
 - Encouragement to slow down and tap out
 - Augmentative communication – objects of reference

Emotional and mental health

- Familiar and predictable and safe environment
 - Physical space, behaviour of others
- Carers with experience/training
- Program of activities
 - Normality, flexibility
- Communication
 - Listening, understanding and respect for individuals, responding to anxieties and worries
- Life story work and memory book/box
 - Bartimeus and Heather House
- Family support

Emotional and Mental Health

- Sleep disturbance
- Anxiety
- Aggressive behaviour
- Depression
- Psychosis and hallucinations
 - related to seizures
 - Related to VI
 - part of disease process

Hallucinations

- Diary and monitoring
 - Triggers, maintaining factors
- Familiar safe, confident, quiet environment
- Attention to lighting, seizure control and activities
- Acute
 - One person, repeating reassurance
 - Respiridone single dose

Drug management Behaviour problems

- Melatonin (Hatonen et al 1999)
- Older antipsychotics
 - Thioridazine
- Newer antipsychotics (Backman et al 2001)
 - Risperidone
 - Intermittent/Continuous
 - Olanzapine
 - Sulpiride
- Pain control

Drug worries

- Lower seizure threshold
- Increased extra pyramidal effects
- Fatigue
- Weight gain
- Not effective
- Malignant neuroleptic syndrome

Bladder and bowels

- Fluid intake
- Varied diet as long as possible
- Active and proactive management of constipation
 - Simple laxatives
 - Stimulants
 - Movicol
 - Enemas as necessary

Feeding and nutrition

- Speech and chewing often together
- Attention to seating and positioning
- Attention to food volume, speed of presentation and textures

- Regular assessment, weights
- Risk of aspiration
- Augmentative feeding – NGT, gastrostomy

Puberty and menstruation

- Acne, hirsutism, polycystic ovaries
 - (Aberg et al 2002)
- ?part of disease process
- ?side effects of AED treatment

- Minimise use of valproate where possible
- Combined oral contraceptive to regulate cycles
- Ovariectomy

Family and Carer support

- Parent support network
- Access to specialist help and advice
- Respite care
- Transitional care
- Research – ability to participate, understand and sometimes fundraise.

Late infantile Batten Disease (NCL) and variants

- Vision
- Seizures
- Mobility
- Communication
- Feeding
- Sleep
- Bladder and bowels
- Irritability

LINCL seizures

- GTCS, Partial, drops
- Myoclonus prominent later
- LI variants may have status, very resistant seizure disorders
- Generally:
 - Valproate, clobazam, topiramate, piracetam helpful
 - Lamotrigine, Carbamazepine unhelpful
 - ? Leviteracetam

Feeding, irritability, bladder and bowels, dribbling and secretions

- Laxatives
- Fluids
- Attention to seating and posture
- Baclofen
- Simple analgesics
- Sleep: melatonin, chloral hydrate
- Hyoscine patch / glycopyrrolate
- Saline nebulisers , physio and suction
- Gastrostomy feeding

Equipment and family/carers

- Housing modifications
- Tracking and hoisting
- Sleep systems and adjustable hospital beds
- Respite
- Parent support network
- Access to experts and research

The UK Batten Interest Group

- Multidisciplinary and interagency
- 3 meetings per year
- Develop clinical guidelines and standards of care
- Share difficult clinical problems
- Increase understanding amongst professionals
- Support and engage in research

Acknowledgements

- Young people and families
- Sarah Kenrick, See-Ability Heather House
- Teaching, nursing, care and therapy staff
 - Dorton House School
 - West of England School for Children with little or no Sight
- Batten Interest Group

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