Putting the ABLE back into childhood disability

Chair: Susan Gibb
Speakers: Catherine Marraffa
         Dinah Reddihough
         Katrina Williams
Outline

• Overview of approaches to disability care, the gaps and opportunities for improvement
• Putting the ABLE back in disability for children with cerebral palsy
• When disability meets child abuse and neglect: a case example
• Questions and Comments please
Different locations or funding for care

• Setting
  – Tertiary/Quaternary hospitals
  – Community Health
  – Disability Services
  – Rehabilitation services

• Type of funding
  – Private sector
  – Public care
  – Non-government organisation
  – Federal
  – State / Territory
Govt details $190 million autism package

Parents of autistic preschoolers will receive up to $12,000 over two years to help pay for early intervention programs.

The assistance is part of the federal government’s election commitment to helping children with autism, outlined on Wednesday.

The package, which includes a rebate of $8,000 in the first two years of early intervention programs, will be available to parents of children aged under six for the next two years to 3,000 families.

Up to 3,000 families would be eligible for the rebate, with a special parliamentary secretary for disabilities Bill Shorten promising, “Some of the behavioural therapies are very expensive, and an early intervention centre, where you’ve got services provided for four hours a day, can cost $30,000 a year.”

Helping Children with Autism

The Australian Government & Helping Children with Autism package recognises the importance of diagnosis and early intervention treatments and services for children with autism spectrum disorder, along with the need for positive partnerships between their families and schools.

The package is being delivered by the departments of Families, Housing, Community Services and Indigenous Affairs (lead agency), Health and Ageing, and Education, Employment and Workplace Relations (DEEWR).

Under the Helping Children with Autism package, DEEWR is responsible for delivering the Positive Partnerships initiative, which aims to build partnerships between schools and families to improve the educational outcomes of children with autism spectrum disorder. Over 2008–12 Positive Partnerships is delivering:

- Professional development for teachers, school leaders and other school staff to build their understanding, skills and expertise in working with children with autism spectrum disorder.
- Workshops and information sessions for parents and carers to assist them to work in partnership with their child’s teachers, school leaders and other staff.

Further information

The Australian Autism Education and Training Consortium (AETCO) is delivering Positive Partnerships workshops across Australia on behalf of DEEWR. More information, including resources for teachers and parents, is available on the Positive Partnerships website.
Different types of services

- Local
- State-based
- Federal

- Early intervention
- Disability support

- Public
- Private
- Non-government sector

- Health
- Education
- Community
- Disability

- Diagnosis based
- Age based
- Needs based

- Preventive
- Primary care
- Secondary care
- Tertiary care
Access varies
Are we developing shared understandings and ways of working?

Are we joining the dots?
Different models and frameworks

• Medical model & Family Centred Care
• Bronfenbrenner’s ecological model
• Social model of disability
• International Classification of Functioning, Disability and Health and the ‘f’ words adaptation
• Economic models
• Human rights – Disability & Child
• Ethical practice
• Evidence-based care
• Chronic & complex care/ Care coordination
• Public health approaches
Social Model

“...... challenge the physical, attitudinal, communication and social environment to accommodate impairment as an expected incident of human diversity.”
Public/Population Health

Specialisation as needed

Secondary care Services for dis- & diff-ability

Primary Care Prevention, identification and early interventions

Discovery
- Biology
- Technology

Minimising impairment

Individual & population-based
Bronfenbrenner's ecological framework
International Classification of Functioning, Disability and Health
Many common themes

Service coordination across artificial silos

Minimise the impairment

Support the carers

Everyone engaged in the community

The built environment

Modify the environment

The way people think and behave

Well being and mental health

Effective individual care
The child and their family’s journey

- In utero
- Perinatal period
- Early childhood
- School years

Is there a problem?

Seeking clarity
- What are the problems?
- Is there a known cause?
- Is there a useful label?
- What does this mean now and in the future?

Accessing effective care and support
- What will help my child?
- What will help me?
- What will help my family?
- Can I access what I need?
- Does our community care?

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The chronological journey

Preschool years
- Identification
- Early intervention
- Parent training
- Parent support

Primary School
- Transition to school
- Education
- Health concerns
- Sibling support
- Parent support

High School and Adult Transition
- Transition to high school
- Working towards independence
- Education
- Work related skills
- Health concerns +/- end of life
- Sibling support
- Parent support
Early Intervention
Federal & State

Carer support

Wheelchair & other equipment

Early childhood

Middle Childhood

Young adult

Mental health services

Hospital Care

Hospital Care

Childcare

Education

Work planning & support

Therapy – speech and language, physiotherapy

Outpatient health care – GP, specialists
Through the Maze

An overview of services and support for parents of children with a disability in Victoria

The “F words”
Thanks to Peter Rosenbaum and Jan Willem Gorter

**Function** – What has your child done well over the past few months?

**Friends** – How is your child getting on with other children?

**Family** – How are you doing? Do you need more support?

**Fun** – Are interventions enjoyable?

**Fitness** – If possible..getting a program to help fitness?
Taking shape
We have the technology

- e records
- Tele-care
Underpinned by and building evidence

Synthesis existing evidence

All services based on best practice

High quality population data

Make best use of existing data
Apply emerging technologies

So we can continually improve

So we can make use of existing knowledge

Ensure evidence translation - service to research and back again
Workforce

• Increasing number of professionals with interest in disability
• Need to understand
  – Commonalities between disabilities
  – Areas where specialisation is needed
• Care coordination expertise is crucial

Unlikely that current training & educational models that are discipline or domain specific will be adequate
National Disability Insurance Scheme

• Insurance model that requires predicted expenditure based on likely outcomes
  – Will need diagnosis and functional assessments
• Being rolled out region by region across Australia

✓ Single entry for services
✓ Registered care providers
✓ Funding for services & products
✓ Comprehensive evaluation
✓ Opportunities for research
Cerebral Palsy
How can we put the ABLE back into childhood disability

The best possible:
- Inclusion
- Participation
- Quality of life for children and their families

Currently some receive good treatment.

For others:
- Diagnosis is made too late
- Families report being poorly informed and supported
- Postural deformities
- Poor nutrition
- Premature death

How can we do better?
The Early Journey

Tom

- Maternal infertility
- Conceived following IVF
- Premature labour at 26 weeks’ gestation
- Many neonatal problems including intraventricular haemorrhage and sepsis
- Some irritability and not sitting at 9 months
What does the paediatrician do?

1. Feels uncomfortable about making the diagnosis/talking about high risk
2. Considers that it is not the right time: let’s wait and see until child is a little older
3. Decides to make a referral to paediatric neurologist so he/she could break the news
4. Believes that early diagnosis/therapy makes no difference
What is the average age of diagnosis of cerebral palsy?

19 months

More severe diagnosed earlier

n=1734 | Australian Cerebral Palsy Register [NSW/ACT] (2011)
So can we change this “late diagnosis”? 

Yes we can.
Early diagnosis can now be made

- General Movements Assessment
  - Good reliability
  - Good validity
- Hammersmith Infant Neurological Examination
  - HINE less than 57 at 3-months 96% predictive of cerebral palsy

2004 – American Academy of Neurology
All children with cerebral palsy of unknown aetiology should undergo neuroimaging, with MRI being the preferred investigation Will confirm neuropathology in about 80%
Early diagnosis

• For infants less than 5 months and at high risk of cerebral palsy
  • Early detection using a combination of General Movements assessment and MRI at near term age over 95% accurate

• For infants older than 5 months corrected age, and at high risk of cerebral palsy
  – Early detection using a combination of the Hammersmith Infant Neurological Examination and early MRI is over 90% accurate
International Clinical Guideline for Early Detection of High Risk of Cerebral Palsy
Does early intervention help?

No firm conclusions

Difficult to generalize because of heterogeneity of interventions, especially type, frequency and duration of interventions and also because of limited group sizes

Suggest that early intervention in the first year of life should consist of a combination of:
- Developmental stimulation that includes trial and error
- Learning in a challenging enriched environment
- Supportive of parent-infant interaction
Conclusion:

• Studies varied considerably in methodological quality, the intervention approaches and outcome measures used and had small sample sizes

  Recommendation for clinical practice are **weak**

• Common themes for the most successful approaches:
  – Child initiated movement
  – Environmental modification/enrichment
  – Task specific training

MORE RESEARCH TO BE DONE: REACH & GAME STUDIES
Many individuals with cerebral palsy die young

Population-based survey 3507 individuals with CP in Victoria. Mortality rates:

• 3% by the age of 5 years,
• 6% by the age of 10 years,
• 11% by the age of 20 years,
• 17% by the age of 30 years,
• 20% by the age of 40 years (Individuals with CP had an 80% survival rate to the age of 40 years)

NO IMPROVEMENT IN SURVIVAL BETWEEN THE 1970S AND 2000S
Can we do better?
Management

• Need to search for the best evidence
• Recent systematic review of 64 discreet interventions for CP, only 24% were demonstrated to be effective, 70% had uncertain effects and 6% were demonstrably ineffective

Management of health issues and related co-morbidities

All children should have:
Hearing assessed
Vision assessed
Be immunised
Have regular dental care

Boys should be screened for undescended testes
Management of health issues and related co-morbidities

Epilepsy

• All seizure types are seen
• Some types of epilepsy are a direct consequence of the underlying brain injury and may persist throughout life.
• Other types of epilepsy are not directly related to a child’s cerebral palsy eg benign childhood epilepsy with centrotemporal spikes (Rolandic epilepsy)
Cerebral palsy and Epilepsy

166 children with CP and WMI
- 41 children (25%) had seizures
  - 30 focal epilepsy typical of childhood epilepsy with centrotemporal spikes
  - 2 idiopathic epilepsy
  - At last follow-up, 80% had not had a seizure for > 2 years
- Favourable outcome in most, important for counselling and drug treatment

63 children with CP and AIS
- 34 children (54%) had seizures, majority were focal seizures suggestive of Rolandic or occipital seizure origin
- Only 15% had active epilepsy 10 years after onset.
Care Pathways - Dystonia

Flow Diagram for an Evidence-Informed Care Pathway for Dystonia in Cerebral Palsy

Assessment
- Determine if dystonia is present (e.g., HAT, neuro exam) and if generalized or focal
- Assess severity of dystonia (history, neuro exam)
- Determine if dystonia is consistent with CP diagnosis (risk factors, brain imaging) or if additional work up is required (e.g., trial of levodopa)
- Assess impact of dystonia on function, quality of life

Determine Intervention Goals for:
- Dystonia causing pain
- Dystonia interfering with function

Introduce Rehabilitation Strategies
- Therapy consultation
- Seating / positioning (pelvic trunk stabilization, hip flexion)
- Assess splinting options (e.g., AFOs)

Generalized Dystonia

Focal Dystonia

Oral Medication Interventions (trial of one or more)

Generalized
- Baclofen (1st Line)
- Trihexyphenidyl* (2nd Line)

Specific
- Painful Dystonia: Gabapentin
- Status Dystonicus: Benzodiazepines & Clonidine
- Dystonia w/ disturbed sleep: Clonidine, +/- Benzodiazepines

Botulinum Toxin

Dystonia is more severe

Intrathecal Baclofen

Deep Brain Simulation

Color Legend for Level of Treatment Evidence
- Effective
- Probably or Possibly Effective
- Data Inadequate (recommendation based on expert opinion)

*Trihexyphenidyl had level ‘C’ evidence of being possibly effective for dystonia; however, continues to be recommended for consideration as a 2nd line oral medication by the care pathway expert team.
Clinical Guideline for Osteoporosis

- **Calcium (Ca) intake**
  - Review dietary intake and determine if Ca intake is adequate:
    - 1–3y=700mg elem Ca
    - 4–8y=1000mg elem Ca
    - 9–18y=1300mg elem Ca
  - Increase dietary intake (1st choice) or consider Ca supplement if required

- **Vitamin D (vitD) supplementation**
  - Consider starting doses of vitD$_2$/D$_3$ between 800-1000 IU

- **Investigations**
  - Check blood work and urine at baseline and aim for:
    - 25-OH-D levels in normal to high range (70–100nmol/L)
    - Ca/Osmolality ratio spot urine in normal range (<0.25)

- **Child/adolescent with osteoporosis (fragility fracture(s))**
  - Consider additional investigations
    - **Lab tests**
      - Calcium
      - Phosphate
      - PTH
      - Alkaline phosphatase
      - 25-OH-D
      - Creatinine
      - Ca/Osmolality ratio (spot urine)
    - **X-rays**
      - X-ray of ‘Symptomatic Area’ to assess for possible fracture
      - Consider need for lateral spine x-ray to assess for the presence of vertebral compression fractures
      - X-ray of wrist for bone age or rachitic changes
    - **DXA scans**
      - Use DXA z-scores adjusted for child’s age, sex, and height

- **Refer for consultation to a bone health specialist**:
  - **Step 1**: Ensure adequate Vitamin D and Calcium
  - **Step 2**: Consider bisphosphonates (use with caution)

- **Colour legend for treatment effectiveness on BMD (evidence):**
  - Effective (A)
  - Probably effective (B)
  - Possibly effective (C)
  - Data inadequate (U)
Clinical Practice Guideline for Sialorrhea

Flow Diagram for Evidence-Informed Clinical Practice Guideline for Sialorrhea in Children/Youth with Cerebral Palsy
Hip Surveillance Guidelines

Effective interventions

General principles

• Goal-directed training
• Child-active structured training in self-care tasks
  – eg dressing, designed so task is meaningful for child
• Involve parent education and encourage parent-child interaction

Therapies shown to be effective

• Bimanual training for hemiplegia
  – 30 – 60 hrs of therapy over 6-8 weeks
• Constraint-induced movement therapy for hemiplegia
  – 30 – 60 hrs of therapy over 6-8 weeks
• Strength training
Improved technology
Parental mental health

294 mothers of children with a disability

High rates of mental ill health

- 44% clinically significant depression
- 42% anxiety
- 21.8% suicidality in the previous 12 months

96% of mothers perceived a need for professional support

- only 54% attempted to access this.

Key barriers to accessing support were

- limited time (44.7%)
- not perceiving the mental health problem as serious enough (36.2%).
Two current research studies

Resourcing Key Workers / Early Childhood Intervention Workers to:

• Detect maternal mental health problems
• Support mothers to access referrals

(Dr Elise Davis, University of Melbourne)
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Putting able back into childhood disability

- Improved evidence
- Use of knowledge that is available
- Greater focus on children within families & their community

TO CREATE A PATHWAY TO A BETTER FUTURE

Put the able back into childhood disability
Disability, Abuse and Neglect
What do we know?
Are children with a disability more likely to live in situations of social disadvantage?

Http://www.abs.gov.au/AUSSTATS/abs@.nsf/Lookup/4102.0Chapter4002008

“Families with at least one young child with a disability have a variety of different experiences and circumstances but tend to have lower socioeconomic status, labour force participation and income than other families with young children. This may be partly due to the larger size of families with a child with a disability.”
What do we know?
Impact of social disadvantage on disability

Predictors of Special Education Resources at Kindergarten Entry
After extreme prematurity

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<td>2.4</td>
<td>1.1 – 5.0</td>
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Msall et al. AJDC 1992; 146:1371-1375
What do we know?

Prevalence of abuse & violence

Systematic review and meta-analysis

Approximately 2000 children under 18 years
17 studies included in final – heterogeneity and variable quality

- Risk of physical and sexual violence three to four times more likely in children with disabilities. Similar rates for neglect and emotional abuse.
- 25% of children with a disability will experience abuse in their lifetime
- did not include all forms of abuse:
  - bullying
  - experience of war or terrorism
  - witnessing domestic violence
Birth

Girl with spina bifida delivered at term
• Born in district general hospital
• No antenatal diagnosis
• Large thoracic level lesion with hydrocephalus and Chiari malformation
• Early transfer to neonatal intensive care, 70km away from home for treatment

Day 1
• Back closed by neurosurgeon and plastic surgeon. Difficult to close
• VP shunt inserted for severe hydrocephalus
Family history

• Second child to mother
• First child to father
• Mother previously worked as an enrolled nurse at closest tertiary institution (30 km)

• 3 year old female sibling - noted to swear a lot when visiting neonatal ward

• Father verbally abused mother on the neonatal ward
Second month of life

- Day 28: baby developed meningitis and CSF leak
- Shunt removed and externalised
- One month later VP shunt reinserted
While hospitalised

- Episode of physical violence in the care be parent unit
- Mother discloses domestic violence and her wish to leave father to social worker
- Abuse - physical and verbal
  - Threats to burn down family home
  - Abduct mother’s older daughter
  - Murder mother

- Father - regular heavy marijuana use and bipolar disorder

- Mother - long standing depression
Post discharge

- Not referred to child protection
- Sent home with early intervention services and with Strengthening Families
- Voluntary

- Mother refused to allow workers into the home
- Failed to attend multiple hospital appointments
- Marked neglect noted by early intervention workers who reported difficulty engaging mother
Increasing services

- Protective services referral made
- Disability services involved
- Mother refused to see one paediatrician in spina bifida clinic following protective services referral
- Transferred to other paediatrician in clinic
- Local paediatrician engaged
- Close liaison

- Parents separate and then reunite multiple occasions
- Next pregnancy – brother born when aged 2 years
School age

- WPPSI performed IQ - 71
- Commences at local primary school with funding for physical disability

Family
- Mother reports difficulties with her other 2 children
- Son - aggressive, hyperactive, constantly demanding
- Older daughter - behavioural problems and learning difficulties
- Maternal mental health deteriorating
Ongoing complex care needs

Education
- Transferred to specialist school aged 10
- Often failed to attend

Mental Health
- Developed marked anxiety symptoms reported by school
Complex health care needs

Neuropathic bowel and bladder
- Catheterised 4 hourly and manual disimpaction by mother for bowel care

Skin care
- Multiple pressure areas
- Weight gain

Orthopaedic problems
- Hip dislocation from dysplasia
- Fractured femur - aged 8 - admitted under orthopaedic unit and no communication with paediatrician
- Kyphoscoliosis - marked deterioration
- Surgery not possible
Complex health care needs

Respiratory problems
- Developed OSA - oxygen requirement
- Severe central apnoea
- Severe restrictive lung disease from severe kyphoscoliosis
- Tonsillo-adenoidectomy aged 10
- On overnight BiPAP at home
- Significant respiratory compromise with frequent presentations to local hospital

PLAN FOR PALLIATIVE CARE DISCUSSED WITH MOTHER
Admitted aged 11

Physical health
- Respiratory infection and UTI
- Severe breakdown of thoraco-lumbar wound with subcutaneous collection
- Head lice

Family breakdown
- Restraining order against father for violence to mother and children
- Breached multiple times by mother and father
- Father remanded in custody
Age 11

Child mental health
- Mental health assessment
- Diagnosis of anxiety and depression made by psychiatrist

Services
- Palliative care involvement led to huge boost in provision of equipment e.g. new bed with ripple mattress, bathroom modifications, extra support with school attendance, respite in hospice
Family situation

- No co-operation from mother
- Multiple attempts to help mother engage with services
- Child protection finally brought a Protection Application by Notice
- One year later agreement that care would be shared with mother and foster carer
- 6 months later, judge asked by child to allow her to live full time with her foster carers
- Granted
Four years later…

• Supervised access with biological mother once a fortnight
• General health improved especially pressure areas and sleep
• Very severe obstructive lung disease but no longer requiring BiPAP and oxygen
• Recurrent UTIs and respiratory infections continue
• Concerns about her mental health continue
• Foster family reporting
  – “Lazy”
  – “Does it on purpose”
  – “Gets angry over nothing”

TRANSITION TO ADULT CARE COMMENCING
A child with a disability born into a situation with high risk of abuse

- End of life an option?
- Family support
- Parenting

Soon after birth

- Family support
- Optimise parental mental health
- Ensure education
- Care coordination

Early Childhood

- Family support
- Young person’s mental health
- Support for the young person to gain independence

Adolescence
An ABLE approach

• Professionals - optimal communication
  – Amongst all professionals
  – With the family

• Well coordinated care

• Relevant support offered

• Parental mental health as a focus
  – Prior mental health issues
  – Secondary to grief and care burden

• Services that are not silos

• “Whole of government” action not rhetoric
Your thoughts?