A Model of Care for Patients with Spina Bifida

Dr C West AM
Spina Bifida Service
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Model of Care for Spina Bifida

1. Neurological
   - Medical: Hydrocephalus, Shunt, Seizures, Vision, Brain stem, Puberty, Tethered cord, Syrinx
   - Functional Learning

2. Orthopaedic
   - Medical: Weakness, Sensory loss, Deformity, Fractures, Pressure areas, Oedema, Scoliosis
   - Functional Mobility

3. Neurogenic Bladder/Bowel
   - Medical: Inefficient emptying, Infection, Renal damage, Stones, Constipation
   - Functional Continence

Lifestyle Impact

- Family: Adjustment, Support, Care Provider, Respite

- School/Education: Learning, Adjustment, Acceptance, Skills development, Independence

- Community Integration: Leisure, Sport, Transport, Relationships, Housing

- General Health: Nutrition, Fitness/Sport, Obesity, Sexuality, Self managed health care

GOAL: Happy, healthy, competent, independent adult.
Explanation of Model of Care

• To provide a framework for holistic care of patient with spina bifida
• Across the age span
• Medical issues, relative importance
• Impairment, relative importance
• Lifestyle issues
• Family/relationship issues
• Health management
One Hundred Complications +

- Medical - Physical
- Medical - Emotional, mental health
- Genetics
- Family/Relationships
- Housing
- Employment
- Finances
Medical - Physical

- Neurological
- Orthopaedic - muscles and bones
- Neurogenic Bladder and renal
- Gastro intestinal and neurogenic bowel
- Skin, soft tissues, sensation
- Back – spinal cord, bones, skin
- Endocrine
Neurological

• Hydrocephalus - shunt dysfunction; disconnection; infection; extravasation
• Meningitis
• Cognitive impairment especially executive functioning
• Arnold-Chiari malformation
• Syringomyelia, Arachnoid cyst spine/brain
Neurological

- Tethered cord syndrome, dermoid cyst, split cord syndrome, diastamatomyelia, arachnoiditis
- Seizures
- Brain stem problems – central apnoea, vocal cord paralysis, swallowing dysfunction
- Eye problems – nystagmus, strabismus, 6th nerve palsy, optic atrophy, amblyopia, impaired vision
Orthopaedic - Muscles

- Lower motor neurone dysfunction - flaccid paralysis of muscles
- Fixed deficit i.e. no improvement in nerve function, no deterioration [unless complications]
- About 10% have some spasticity in lower limbs
- Sensory deficit limits feedback about muscle function, position of limb, learning to walk
Orthopaedic - Bones

- Joint Deformity, dislocation, subluxation
- Fractures
- Osteoporosis, osteopenia
- Joint deformity, instability
- Osteomyelitis
- Bursa
- Charchot’s joint
- Osteoarthritis
Neurogenic Bladder / Renal Complications

- Inefficient emptying of bladder
- Infection of urine
- High pressure bladder
- Ureteric reflux
- Hydronephrosis
- Renal damage – renal failure
- Calculi - bladder, kidneys
- Hypertension
- Incontinence
Neurogenic Bladder- Intervention Complications

- Ureteric stricture
- Conduit - stenosis, stretching, calculi
- Vesicostomy – stenosis, dermatitis
- Augmentation – hyperchloremic acidosis, calculi, rupture, metaplasia
- Appendicovesicostomy – stenosis, granulation
- Dialysis - multiple issues
- Renal transplant – multiple issues
Gastrointestinal Complications

- Constipation/Diarrhoea
- Incontinence of bowel
- Peritoneal shunt
- Inguinal hernia
- Rectal/vaginal prolapse
- Haemorrhoids/anal fissure
- Gastro-oesophageal reflux
Gastrointestinal – Intervention Complications

- Peritonitis - shunt infection
- Adhesions – bowel obstruction
- Bladder augmentation – bladder rupture
- Caecostomy button/stoma – stenosis, abscess
- Caesarean Section – following abdominal surgeries
- Appendicitis/ pelvic abscess – late presentation reduced pain
Skin and Soft Tissues

- Sensory loss – reduced pain, position sense
- Pressure areas
- Cellulitis
- Ulcers
- Sinus tracts/ fistulas
- Chilblains
- Poor circulation/discoloration/cold limbs
- Excoriation [due to incontinence]
- Latex allergy
Skin and Soft Tissues

- Lymphoedema
- Deep Venous Thrombosis
- Burns
- Capillary Haemangioma in Myelomeningocele repair area
- Hairy patch in MM repair area
- Excoriation/skin breakdown in MM area secondary to scoliosis/lordosis
Structural Spinal Complications

- Back pain – multiple causes
- Scoliosis
- Kyphosis
- Lordosis
- Hemivertebrae/block vertebrae
- Pelvic obliquity
- Postural and seating issues
- Disc degeneration
Respiratory Complications

- Central apnoea
- Obstructive apnoea [e.g. vocal cord paresis]
- Sleep apnoea
- Restrictive lung disease due to scoliosis/kyphosis
- Pneumonia
- Use of CPAP/BIPAP
Endocrine Complications

- Short stature
- Reduced growth hormone/ slow weight gain
- Precocious puberty
- Delayed puberty
- Male infertility
- Undescended testes
- Breast development [disproportionate to short spine]
- Obesity
Aetiology and Genetics

• Family History
• Syndromes – e.g. Waardenburgs, trisomy 13 or 15, cloacal anomalies, imperforate anus
• Dietary/drugs/alcohol/foetal alcohol syndrome/anorexia
• Risks for procreation/use of folate – patient, other family members
Mental Health Issues

• Adjustment disorders – family, patient
• Anxiety
• Depression – parent’s, patient
• Dementia - ? Earlier onset
Family Complications

• Adjustment  -  Parents, siblings, other family members, patient growing up
• Time commitments to caring
• Hospitalisations – planned, urgent
• Sibling concerns
• Financial impacts
• Future children
Housing

• Secure affordable housing
• Accessible housing
• Access to medical care and education
• Changing needs as child grows
• Transport from home to school, medical care, employment, leisure activities
Education and Employment

- Maximising education close to home
- Extra support for school work
- Planning realistic future goals
- Employment – physical AND cognitive abilities
- Work efficacy may be limited
- Transport to work/driving/costs
- Illness/hospitalisation – impact on work
Relationships

- Within the family
- School friends – building/keeping relationships
- Sexuality/sexual behaviour/vulnerability
- Sexual functioning
- Marriage expectations for both partners
- Reproduction/fertility
- Parenting
Planning/Managing Pregnancy

• Relationships – reality/practical basis
• Contraception
• Motivation for pregnancy
• Genetics
• Maternal health
• Ability to be a parent
• Management of pregnancy/mobility issues
• Management of delivery
• Support for baby care – physical/emotional
• Problem solving re baby’s care needs
Finances

• Financial implications for family – mothers ability to work /child’s needs
• Budgeting for patient
• Dealing with Centrelink/DSP/part time work
• Equipment/housing/transport costs
• Access to subsides and other assistance