

A Model of Care for Patients with Spina Bifida

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Model of Care for Spina Bifida

1. Neurological

- Medical**
- Hydrocephalus
 - Shunt
 - Seizures
 - Vision
 - Brain stem
 - Puberty
 - Tethered cord
 - Syring

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

Neonatal



Pre School



Primary School



High School



Adult



Lifestyle Impact

Neonatal
Preschool
Primary School
High School
Adult

- Family**
- Adjustment
 - Support
 - Care Provider
 - Respite

Neonatal
Preschool
Primary School
High School
Adult

- School/Education**
- Learning
 - Adjustment
 - Acceptance
 - Skills development
 - Independence

Neonatal
Preschool
Primary School
High School
Adult

- Community Integration**
- Leisure
 - Sport
 - Transport
 - Relationships
 - Housing

Neonatal
Preschool
Primary School
High School
Adult

- 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
- Charcot'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 subsidies and other assistance

