

Spina Bifida – Meningomyelocele

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About Neural Tube defects

The incidence of congenital anomalies that manifest at birth is 6% worldwide. Neural Tube defects are some of the most disastrous birth defects and often affect lower limbs as well as urinary tract, which further culminate into life threatening renal disease.

The reported incidence of Neural Tube defects in Europe is 0.7 in 1000 live births, while that in the US is about 0.9 in 1000 births. In India the estimated rate of Neural Tube defects is 4.1 per 1000 live births, which is far higher than the incidence in the developed world. Considering the 26 Million babies born in India every year, around 1.07 Lakh babies are born with Neural Tube defects in India.

This is indeed a staggering figure. The high incidence of Neural Tube defects in India is probably related to the deficiency of Folic Acid in pregnancy. Although the incidence of Folate deficiency in on the decline with the increasing awareness, the incidence of these defects remains alarmingly high. Consanguinity of marriages does play an important role in the higher incidence of such

malformations in our society. Non-detection of birth defects and stringent laws about termination of pregnancy may add difficulties in reducing this number.

The Antenatal Aspects of Neural Tube defects

Neural Tube defects can be picked up by standard Ultrasound antenatally by 12th week of gestation. This is possible due to identification of the neural ridge and the size of the fourth ventricle in the foetus. By about 19 weeks of gestation, defects in the spinal column and herniation of the meninges can be picked up by ultrasound in 95% of cases.

However the presence of neural tissue in the meningeal sac may not be diagnosed with certainty at this time. The Legal Cell of Government of India allows termination of pregnancy of up to 20 weeks, with proven serious foetal malformations. After this period, a special court has to sanction the permission for medical termination, after it is ultimately approved by the local Government bodies.

Besides the incidence of Myelomeningocele (herniation of contents of the spinal cord into the prolapsed meninges) remains fairly high, leading to the affection of the lower limbs as well as the urinary tract.

Factors that influence Myelomeningocele

1. The bony level of the defect.
2. Deferring growth rates of the bony skeleton and the nervous tissue.
3. Development of Arnold Chiari malformation (Protrusion of Cerebellar tonsils through the Foramen Magnum and subsequent compression of the fourth ventricle producing Hydrocephalus)
4. This compression can also affect the function of the Pontine Mesencephalic Centre that controls the function of the bladder.

Evaluation of Spinal Dysraphism

Evaluation includes a thorough Clinical examination with special reference to neurological examination. Sensory and Motor function of the lower limbs must be documented carefully. A very careful examination for associated congenital malformations involves documentation of muscle wasting, equinovarus deformities of foot as well as claw toes. Note the trophic changes of the skin.

A special mention must be made of ammoniacal dermatitis in the perigenital region in both sexes, which indicates long standing urinary incontinence.

Physical Examination should include documenting the integrity of the sacral reflex arc (SRA), which includes estimation of Anal Tone, Perianal sensations, Superficial Anal reflex and Bulbocavernosus reflex.

Laboratory evaluation includes Haemogram, Serum Creatinine level and Urine Culture as baseline. A plain X-ray of the lumbosacral spine is useful adjunct to Ultrasound examination of the entire urinary tract. X-ray should document the bony spinal defect and a bladder shadow. Ultrasound should document renal dilatation, calculi, ureteric anatomy, bladder capacity, bladder outline, presence of bladder diverticula, stones and residual urine volume. They form a baseline for the future reference.

Cystoscopy is generally not necessary for evaluation, unless abnormalities such as diverticula and calculi are demonstrated in the previous evaluation.

By the time these patients are referred to the Urologists, they would have had closure of the Meningomyelocele by a Neurosurgeon soon after the birth. They would also have had some Orthopedic corrections in the feet and other parts of the lower limbs. Usually an initial MRI of the spine is available.

Is Urodynamics mandatory?

Urodynamics evaluation is absolutely mandatory as a pre-emptive and baseline investigation in all cases of spinal Dysraphism. The first evaluation is recommended in the first three months of life, although neurological status may continue to change in the later life. Baseline and follow-up Urodynamics evaluation is necessary in all cases of Myelomeningocele like no other disorders. Addition of a video study may enhance initial evaluation but might not be a part of follow-up Urodynamics evaluation. The main object of this evaluation is:

1. Baseline morphology is documented.
2. Serial monitoring shall estimate the risk to the upper urinary tracts.
3. Helps counselling of the parents
 - 20% have normal Urodynamics
 - 63% show Neurogenic Detrusor Overactivity

Documented combinations of Urodynamics patterns

1.	Synergic Sphincter	20%
2.	Dyssynergia (with or without low Compliance)	37%
3.	Sphincter Denervation	36%

- 71% who have Detrusor Sphincter Dyssynergia, show upper tract deterioration in first three years of life.
- Detrusor Compliance seems worse with patients of high outlet resistance.
- Compliance may improve after "outlet dilatation."
- 20% have abnormal urinary tract on radiology when first evaluated.

Management

Management of Meningomyelocele patients with neurological affection have the following priorities of management.

1. Prevention of Renal Damage
2. Control of Urinary Incontinence
3. Restoration of Bowel Dysfunction
4. Sexual Rehabilitation
5. Psychological Rehabilitation

Urinary Tract always that takes the toll of the systemic damage.

Storage function of the urinary bladder.

Kidneys manage to clear the blood of all chemical impurities and form 500 to 1500 mls of urine every 24 hours. This liquid has to be stored safely for periodic expulsion for social reasons. Urinary bladder therefore has following functions to perform.

1. Store urine at “Low pressure” in order to allow antegrade propulsion of urine from the kidneys round the clock
2. Maintain continence during the storage phase.
3. Prevent reflux of urine upstream into the kidneys
4. Prevent any biochemical change in the urinary constituents during storage, unlike other hollow organs in the body
5. Execute efficient and complete expulsion of urine during voiding

These functions are accurately executed through the precise neurourological axis, consisting of Cerebrum, midbrain, Pons, spinal cord and the pelvic nerve plexus that work in a coordinated fashion and synchronously with the pelvic floor and the external urinary sphincter.

Principles of management for any Neuropathic Bladder

1. Maintain bladder pressure below 20 cms of H₂O at physiological volumes.
2. Note the upper tract changes in patients of voiding detrusor pressures in excess of 25 cms of H₂O.
3. Keep the bladder Residual volumes as low as possible.

Management modalities

1. ISC (or CIC), along with drugs (Anticholinergic and /or Beta-adrenergic) may prevent upper tract damage by keeping the storage pressures to low pressure zone.
2. Refractory individuals may respond better to Intradetrusor (suburothelial) injection of Onabotulinum Toxin.
3. Neurologic features continue to change with age. Hence, sequential Urodynamics evaluation must be performed every year till 5 years of age. Subsequent evaluation should be performed as and when the new neurological signs develop or if there is progressive incontinence or hydronephrosis.
4. Indications for surgical or injection therapy for vesicoureteric reflux are no different than for patients with normal bladder function.
5. Management by CIC and drugs reduce the incidence of vesicoureteric reflux by 40%
6. Crede voiding by suprapubic compression can induce reflex Sphincter contraction and worsen the voiding efficiency.
7. Augmentation Cystoplasty (with Mitrofanoff) is naturally indicated for exceptional cases, which are refractory to conventional management and in patients who show progressive signs of high storage pressures, low capacity, upper tract dilatation and refractory incontinence.
8. Xiao Microanastomosis of L5 ventral root to S3 ventral root has had limited but promising success in restoring the bladder function.
9. Unilateral TUU and use of distal ureteric stump as Mitrofanoff Catheterisation Conduit has had encouraging results.

10. Autologous sling and CIC to control incontinence is a promising option, although its effect on storage function of the bladder has not been documented.

Meningomyelocele in India

In summary, neural tube defects and in particular, Meningomyelocele adds a very heavy burden on the family (and in turn on the society) in terms of short and long term management in India for following reasons.

1. High Incidence of Neural Tube Defects in our population.
2. Higher incidence in the lower socioeconomic group.
3. Poor facilities of antenatal diagnosis of this condition.
4. Stringent rules about termination of Pregnancy*
5. Massive ignorance about potential correction of the problems in the general population
6. Lack of awareness among the primary care physicians that such a condition needs early evaluation and management
7. Associated morbidities like poor mobility, trophic ulcers, pressure sores and faecal incontinence.
8. Associated orthopaedic deformities and ophthalmic abnormalities
9. Social stigma.
10. Psychological consequences.

India's specific Myelomeningocele related needs

1. Promote social awareness of this debilitating and potentially fatal condition.
2. Improve nutrition and Folic Acid supplements among pregnant women
3. Encourage awareness about early Antenatal Ultrasound examination to diagnose this condition.
4. Allow freedom to termination of pregnancy on the diagnosis of severe forms of this condition.
5. Once born, we need a prompt evaluation by a standardised protocol of investigations and management.
6. Promote awareness among Primary care Physicians (as well as Obstetricians and Neonatologists) about the need to commence early evaluation and management by a standard protocol.
7. Encourage multidisciplinary actions among Urologists, Neurosurgeons, Neurophysicians and Orthopaedic Surgeons to rehabilitate this group of patients.
8. Physiotherapists, Rehabilitation Specialists, Incontinence advisors and Psychotherapists must perform subsequent rehabilitation.
9. Once they reach puberty, they need a sexual rehabilitation with the help of Andrologists and sexual counsellors.
10. Finally it is the multidisciplinary responsibility where all subspecialties must understand their roles as well as mutual responsibility in order to give these unfortunate children a decent quality of life, which is immensely feasible.

Antenatal scans timelines

6 Weeks	Viability of the foetus
12 weeks NT scan	Neural Tube defects can be picked up
20 weeks	Almost all anomalies can be diagnosed.

Sexuality

- 72% of male children are able to have penile erection.
- 67% male children are able to ejaculate.
- Men are more likely to get affected.
- Girls reach puberty earlier since there could be Hydrocephalus induced changes on the Pituitary function.
- Bowel function are unpredictable.
- ACE (Antegrade Continence Enema) using appendix or button Caecostomy can cleanse the bowel every alternate day.

Occult Spina Bifida

This is a group of conditions that affect formation of the spinal column but do not result in open vertebral canal. This includes Tuft, Lipoma, skin dimple, vascular malformation etc. There is clawing and arching of the foot, muscle wasting of the lower limbs. They may have normal neurological examination but almost always have abnormal Urodynamic behaviour.

There is disparity in growth spurts between vertebral column and neural elements. 70% have UMN lesions and 30% have LMN lesions. Posterior Tibial Somatosensory evoked potentials are more sensitive indicators of cord tethering on urodynamics.

Such patients must be operated before the age of 3 years. The procedures could include Laminectomy, which is the removal of intraspinal processes.