

Medi Quest BRS Hospital

A monthly News letter from BRS Hospital

NEUROFIBROMATOSIS SCOLIOSIS

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Price Rs. 5/- Only

August- 2023

Medi - 18

Quest - 8

Yearly Subscription

Rs 50/- only

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Neurofibromatosis is a genetic disorder characterized by tumors or neurofibromas that grow in the nervous system and under the skin. The disease occurs as two distinct types -

Neurofibromatosis 1 (NF1) and Neurofibromatosis 2 (NF2) with each type having a unique set of manifestations. NF1 occurs more commonly, in approximately 1 in 3000 births, whereas NF2 affects roughly 1 in 25,000 individuals.

Children with NF1 frequently develop orthopaedic problems. The two most common are spine deformities, particularly **scoliosis** - a curvature of the spine in a sideways or lateral orientation - and **tibial dysplasia**.

The spinal deformities in Neurofibromatosis scoliosis are severe and are often associated with spinal cord compression with

paralysis of the upper and lower limbs depending on the level of involvement of spine. These deformities are difficult to treat requiring specialized surgical equipments and techniques.

Tibial Dysplasia refers to failure of the tibia to develop appropriately. Patients with tibial dysplasia may also have a tendency to fracture and to develop pseudarthrosis, which refers to failure of the fracture to heal properly. Tibial dysplasia can also result in limb length discrepancy, where one leg is longer than the other. Other children with NF1 may develop overgrowth of a limb without any underlying bony abnormalities; in these cases, the underlying problem is soft tissue overgrowth.

In contrast to NF1, NF2 is associated with “acoustic neuromas” that may affect hearing and balance and frequently do not have any orthopaedic manifestations of their disease.

Surgery

Neurofibromatosis scoliosis most often progresses with time. Hence a scoliosis surgery is recommended to reduce the severity of the spinal curve and to prevent it from getting worse. The most common type of scoliosis surgery is called spinal fusion. Surgery to correct a neurofibromatosis scoliosis curve can present special challenges, including the need to correct a kyphotic curve at the same time. Halo vest distraction a method of reducing the severity of deformity before fusion surgery is used in certain situations.

In spinal fusion surgery, surgeons connect two or more of the bones in the spine (vertebrae) together, so that they cannot move independently. Pieces of bone (bone graft) or a bone-like material are placed between the vertebrae. Metal rods, hooks, screws or wires typically hold that part of the spine straight and still while the vertebrae fuses together.

While there is no cure for neurofibromatosis, careful monitoring and intervention for orthopedic and other complications can help improve the patient's quality of life significantly.

ADVANCED SURGERY FOR COMPLEX SPINAL DEFORMITY

A 19 years old student from Elampillai, Salem District was admitted with complaints of inability to walk since 3 months in the Ortho Spine Surgery Unit of Institute of Orthopaedics and Traumatology, Rajiv Gandhi Government General Hospital, Chennai. Clinical examination revealed deformity of the upper thoracic spine (Left sided kyphoscoliosis) which was very severe (sharp kyphosis). Neurological examination revealed paralysis of both lower limbs with complete loss of sensation. The presence of skin patches (café au lait spots) helped in making a diagnosis of Neurofibromatosis Kyphoscoliosis.

The team headed by **Prof. Nalli R Uvaraj, Chief Ortho Spine Surgeon, Madras Medical College and Rajiv Gandhi Govt. General Hospital, Chennai and Orthopaedic Consultant, BRS Hospital, Chennai** applied a Halovest traction and distraction of the

deformed spine was performed daily. This was carried out for a period of 6 weeks. With this procedure patient showed some neurological recovery . At a Second Stage patient was taken up for Posterior stabilization and spinal fusion. Special screws and rods were used to correct deformity and stabilize the spine. Bone graft was applied to fuse the spine.

Subsequent to the surgery patient showed further improvement of power and sensation in the lower limbs. She started walking with support during her stay in the hospital. She was discharged with the advice to continue physiotherapy.

At her last follow-up the patient had complete recovery of motor power and sensation in the lower limbs. She at present walks independently and is planning to join college to continue her studies.





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Owned and Published by Dr. Madhusudhan 28, Cathedral Garden Road, Chennai - 34.

Printed by S. Baktha at Dhevi Suganth Printers 52, Jani Batcha Lane, Royapettah, Chennai -14.

Publication on : Final Week of Every Month Posted on 31.0.2023