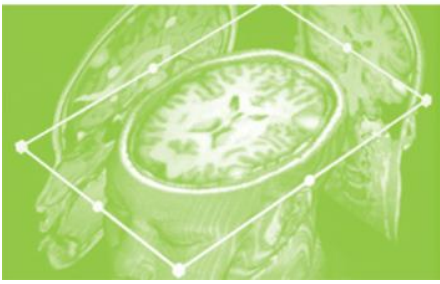
Children's
Healthcare of Atlanta

Volume 2, Issue 1



Neuro Update℠

NEURO SPINE CARE FOR KIDS

David Wrubel, M.D., and Barun Brahma, M.D.

When the spine is properly functioning, a child can run, jump, play and learn—a kid can be a kid. However, when the spine malfunctions due to a birth defect, tumor, injury or disease, a child's activity level can change overnight.

The Pediatric Spine: Not Just an Adult Spine

To use an old adage, children are not small adults. This statement is particularly appropriate when discussing the spine. At birth, the newborn spine is relatively straight, lacking the normal sagittal curves seen in adults. When the newborn begins holding his head up, the cervical lordosis develops, which is followed by development of the lumbar lordosis when the child begins standing and walking. A child's head proportionately accounts for far more total body area and mass than an adult's head, a roughly one-to-four ratio in infants compared to a one-to-seven ratio in adults.

This, combined with disproportionately weak cervical musculature and small spinous processes, places greater stress on the pediatric cervical spine, helping to explain why the cervical spine is the most injured segment of the immature spine. Other significant differences between the pediatric and adult spine include anterior wedging of the vertebral bodies; more horizontal orientation of the facet joints; and overall laxity of the spinal ligaments (particularly the interspinous ligament). These factors both predispose children to a different pattern of injury and disease and present special challenges in the management of pediatric spine pathology.

Back and Neck Pain
Back and neck pain are the most common complaints to pediatric neurological spine surgeons. The majority of these complaints are related to over-exertion, minor trauma and the day-to-day activities of active children. Rest, over-the-counter analgesics and time are the mainstays of therapy, and most cases resolve within several days. If pain persists beyond this period of time, it merits further consideration. The features suggesting a problem include pain that does not respond to over-the-counter analgesics; pain radiating down the arm or leg (radiculopathy); weakness and/or numbness; change in bowel or bladder function; focal pain in the midline spine; pain that does not improve or worsens when lying flat (recumbent pain); or worsening pain despite conservative management. Any of these symptoms should prompt further investigation, as more significant pathology is likely to be found.

Basilar Invagination and Basilar Impression
In children with these rare craniocervical junction disorders, the skull progressively settles down on the upper cervical spine to the point that the spine is pushing up into the brainstem. Basilar invagination (primary invagination of the spine through the foramen magnum) and basilar impression (invagination of the spine through the foramen magnum secondary to a separate primary disease process, such as Hajdu-Cheney syndrome) are slow, progressive processes and lead to a change in voice, dysphagia, respiratory symptoms, gait changes, myelopathy and even death. Traction to reduce the displaced spine and/or transoral resection of the odontoid combined with occipitocervical fusion can halt progression and often reverse the progressive brainstem and cranial nerve dysfunction.

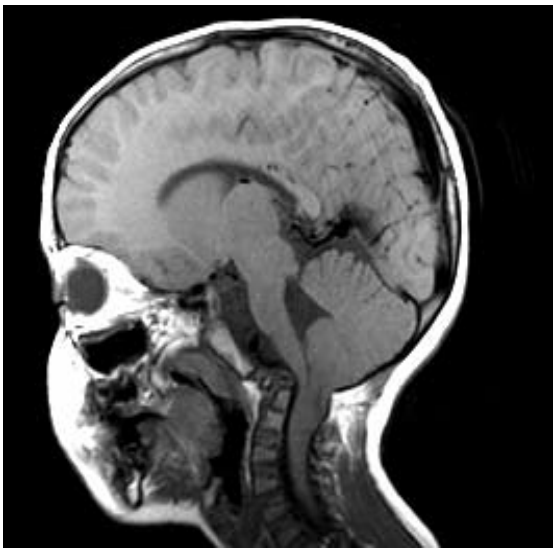


Figure 1: 5-year-old child with basilar impression. Note the compression of the brainstem from the dens.

Trauma
Injuries to the spine include conditions from simple muscle strain to complete paralysis. In addition to the immediate effects of spinal injury, delayed problems, such as instability, scoliosis, kyphosis and chronic pain, can develop years after the initial injury. Due to the specific properties of the immature spine, patterns of injury are different at different ages, and several injuries, such as spinal cord injury without radiographic abnormality, are only seen in children.

Spine Tumors
Spinal cord tumors commonly present as back pain, often occurring at night while in bed (recumbent pain), and/or progressive neurological deficits, such as hemiparesis, hypesthesia and ataxia. Certain conditions, such as neurofibromatosis, predispose children to the development of spinal cord tumors, but the majority arises in otherwise healthy children.

Treatment almost invariably involves microsurgical resection with, or without, postoperative adjuvant therapy. Tumors arising from non-neural tissue that impinge on the spinal cord include neuroblastoma, sarcomas, teratomas and metastases. These tumors can invade and destroy the boney spine on their way to compressing the spinal cord and nerve roots, or they simply invade the spinal canal via the neural foramina. Extensive resection, with, or without, spinal reconstruction, is often required as is postoperative adjuvant treatment. Tumors of the boney spine vary from benign, incidentally found lesions, to destructive malignant neoplasms compressing the spinal cord. Pain is the most common initial symptom, and it can be dramatic. Equally dramatic can be the pain relief afforded by a single aspirin, a provocative and diagnostically suggestive feature of certain spinal column tumors. Accurate diagnosis is critical, as some of these tumors should be conservatively followed, while others require extensive resection and spinal reconstruction.

Figures 2A and 2B: Flexion/extension cervical spine series in a 6-year-old girl with Down syndrome and atlantoaxial instability.

Craniocervical Junction Disorders
Disorders of the craniocervical junction include congenital abnormalities (segmentation defects, assimilation abnormalities, hypoplasia or absence of spinal elements and congenital fusions) and developmental and acquired abnormalities (achondroplastic stenosis, osteogenesis imperfecta and renal rickets).

In addition to neck pain, disorders of the craniocervical junction present with symptoms related to compression of the lower brainstem and upper cervical spinal cord: dysphagia, dysarthria, myelopathy and spasticity. Other signs and symptoms include torticollis, respiratory compromise/sleep apnea, headache and an abnormal appearance of the neck. Occult craniocervical junction abnormalities often become apparent after a deceptively minor traumatic event renders a child neurologically impaired. If there is instability, the key issue with these disorders is determining the stability of the abnormal region and preventing neurological injury by surgically fusing unstable segments. In the absence of instability, aggressive, nonoperative management—headed by Children's Healthcare of Atlanta rehabilitation and pain management specialists—controls pain and helps improve a patient's quality of life.

Atlantoaxial Instability
The first two cervical vertebrae are unlike any other in the spine. Apart from the direct articulation of the atlas and skull, the characteristic articulation of C1 and C2, absence of a disc space, the odontoid process and the unique ligamentous structures of this region make it particularly vulnerable to injury and progressive pathology. Because of the relatively greater mass of the pediatric head, the cervical flexion fulcrum lies closer to the C1 to C2 region in children than it does in adults—further predisposing this area to injury and developmental instability.

Conditions and diseases associated with atlantoaxial instability include Down syndrome, Morquio's syndrome, juvenile rheumatoid arthritis, os odontoideum and trauma. Abnormal movement in this region can lead to progressive myelopathy, spinal cord injury or ventilator- dependent quadriplegia, if severe. Management determines the degree of instability and the possibility of surgical fusing of the C1 to C2, depending on if the degree of instability caused the injury or if the degree of instability puts the child at undue risk for spinal cord injury.

Herniated Disc
Disc herniation in children is becoming more common, and the presentation, management and outcome of this pathology significantly differs from adults. Back pain is a far more common presenting symptom among children than with adults, as radiculopathy tends to be the overriding pain feature. Children, unlike adults, tend to have large, broad-based and centrally located herniations, as opposed to the classic focal lateral fragment seen in adults.

In our experience, children who have no response to conservative therapy after six to eight weeks are unlikely to ever do so. Microsurgical discectomy outcomes in children are better than those reported by adult literature, with nearly 100 percent patient satisfaction and very low complication rates. More importantly, children return to school and activities significantly faster after surgical treatment of refractory disc disease than after prolonged conservative management.

Spondylolisthesis
Spondylolisthesis is a condition of the spinal column where one vertebra is displaced with respect to the next. The most common form of spondylolisthesis in children is isthmic spondylolisthesis, often called a pars defect.

The pars interarticularis is a distinct segment of the posterior-lateral vertebra roughly between the lamina and pedicle. Whether due to a congenital defect or trauma (most commonly a stress fracture), the pars disruption can lead to back pain, progressive instability of the spinal column and, in some cases, spinal cord compression. Laminectomy followed by spinal realignment and fusion is dramatically effective in relieving pain; decompressing the nerve roots and/or spinal cord; and correcting the deformity. The majority of children who are tracked over time with conservative treatment of consistent pain medication and physical therapy will improve without needing an operation. Operative intervention is reserved for the continued progression of their symptoms and/or spondylolisthesis. In most cases, there is no reason to restrict activities during this period, including athletics.

Chronic Pain
Back and neck pain is the most common reason for visits to adult primary care providers, and is becoming a growing problem in children. While the majority of children with back and neck pains have self-limiting muscular strain, chronic pain due to a variety of neurological and musculoskeletal conditions can be a life-altering problem. Additionally, back and neck pain can be the first sign of a serious condition, such as a spinal cord tumor, occult spinal instability and multiple sclerosis. Evaluation of back and neck pain is complex and includes radiological imaging, neurological testing, psychological testing and diagnostic injections.

Treatment is based on the etiology of the pain and may involve medications, physical therapy, therapeutic injections or surgical treatment of distinct pathology (discectomy, fusion and resection of mass lesions). Children with chronic pain due to spinal cord injury or prior surgery can benefit from implantation of a spinal cord (dorsal column) stimulator to interrupt the ascending pain pathway. Finally, chronic pain from certain systemic malignancies can be effectively treated by ablative spinal cord procedures to permanently abolish the involved pain pathway, which greatly improves the patient's quality of life.

Hyperhidrosis
Sweating is a useful physiological process in response to excessive body temperature. When this response is inappropriate or exaggerated, sweating can become life-altering. Hyperhidrosis is a primary sweating disorder mediated by the sympathetic nervous system. Children with this disorder excessively

and continuously sweat, primarily on the palms, soles and axillae. Excessive sweating occurs irrespective of room or body temperature, anxiety or obesity. Hyperhidrosis makes tasks, such as writing, driving and athletics, difficult—if not impossible.

In addition to the functional limitations imposed by this disorder, children with hyperhidrosis feel stigmatized, becoming socially withdrawn. The first-line treatment for this disorder uses topical agents, such as Drysol, in an attempt to control the sweating. Quite often, topical agents and other medical treatments fail to control sweating. In which case, a permanent surgical treatment can be used. Thoracoscopic sympathectomy, a minimally invasive procedure in which the sympathetic nerves to the extremities are severed, typically provides immediate, permanent cessation of excessive sweating.

Fig. 3A: Postoperative lateral cervical spine film showing technique of C1 lateral mass screws and C2 pedicle screws in a 6-year-old child with an os odontoideum.

Fig. 3B: Postoperative axial CT test showing C1 lateral mass screws.

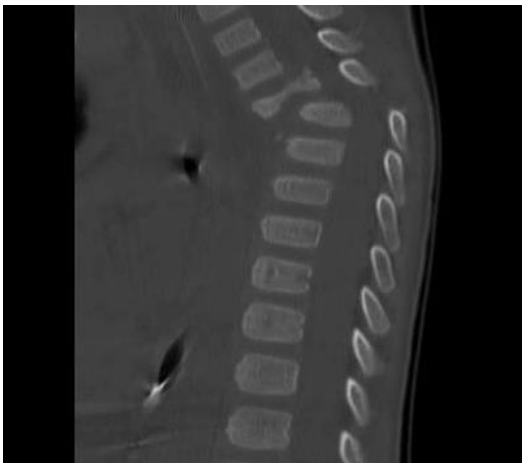


Fig. 4A: CT test of a 2-year-old child involved In a motor vehicle accident with a T4 fracture dislocation.

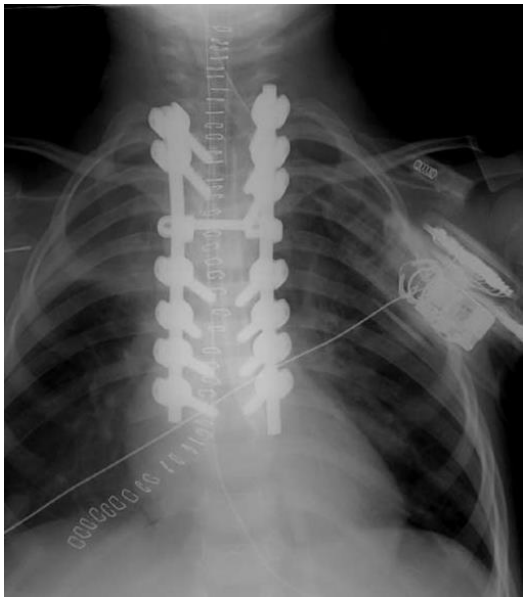


Fig. 4B: Postoperative film showing anteroposterior view of multiple pedicle screw and rod fixation of fracture.

Neuro Spine Care for Kids
The Children's Neuro Spine Care for Kids program has a team of physicians, therapists and nurses dedicated to the evaluation and management of pediatric spinal disorders from diagnosis through rehabilitation. The team includes neurosurgery, pain management, rehabilitation medicine, orthotics, physical therapy, neuroradiology and oncology.

Because children deserve physicians and therapists who know children, every member of the Children's Neuro Spine Care for Kids program is specially trained in pediatrics, caring for children and adolescents through the age of 21. As a true multidisciplinary clinic, each patient is thoroughly evaluated by the appropriate team members prior to a team meeting, during which each child is discussed. A group evaluation and treatment plan is then presented to the family, typically the same day. The child and family are then guided through child-specific, multidisciplinary treatment (surgical and nonsurgical), rehabilitation, pain management and long-term aftercare.

The Children's Neuro Spine Care for Kids program is at the forefront of modern spinal surgery employing the use of intra-operative neurophysiologic monitoring, pediatric-specific spinal instrumentation and techniques, stereotactic intra- operative spinal navigation and minimization of external fixation devices (halos).

Our team is never satisfied with the current state of technology and treatments. So, the Children's Neuro Spine Care for Kids program is involved in research efforts to better understand and treat these complex disorders, including the development of pediatric-specific equipment, instrumentation and techniques.

The final component of the Children's Neuro Spine Care for Kids program involves educating children, families and pediatric care providers about spinal disorders from prevention to treatment options through conferences, symposiums, pediatric-specific literature and, most importantly, one-on-one time to discuss the issues each child faces.

To provide care for children requiring a multidisciplinary approach to their spinal disorder, a team member is available 24 hours a day to answer questions or facilitate a formal evaluation. For children living outside of metro Atlanta, the Children's Neuro Spine Care for Kids program will help families with travel arrangements and local housing for the length of their stay in Atlanta. Emergency referrals can be made 24 hours a day by speaking directly with one of our neurosurgeons who can rapidly arrange either ground or air transportation.

Fig. 5: Coronal CT reformat of a C4 osteochondroma in a 5-year-old boy with a mass growing in his neck.

Fig. 6 T5 metastasis from a parietal lobe pleomorphic xanthoastrocytoma in a 16-year-old girl. The patient only presented with back pain and underwent complete resection of the tumor.

Fig. 7: Magentic resonance imaging (MRI) showing traumatic L4 to L5 spondylolysis with disruption of the intervertebral disc at L4 to L5.

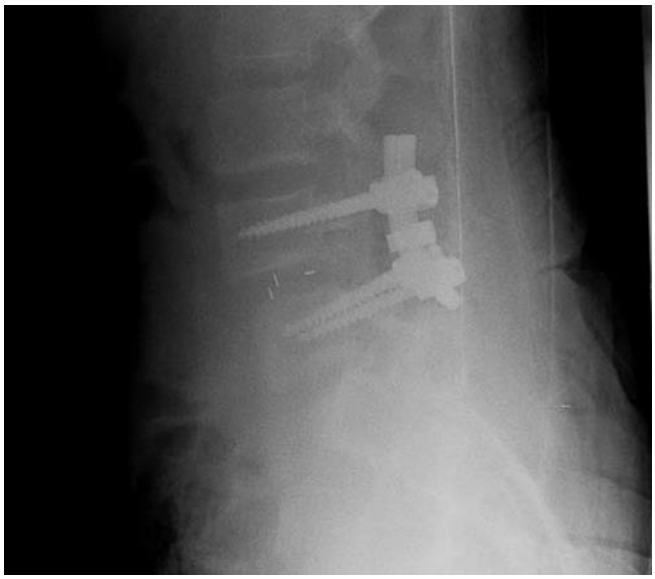


Fig. 7A: Postoperative imaging showing pedicle screw and rod fixation at L4 to L5 with resection of L4 to L5 intervertebral disc and insertion of cage.