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Intraoperative Sonography in Spinal Dysraphism and Syringohydromyelia

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The use of intraoperative sonography was analyzed in 24 patients with spinal dysraphism and syringohydromyelia in order to determine the role of real-time sonography in the surgical management of these patients. Specific diagnoses included tethered cord (nine), syringohydromyelia (seven), congenital tumor unassociated with a tethered cord (four), diastematomyelia (three), and occult sacral meningocele (one). Intraoperative sonography determined the exact relationship of congenital tumors to the cord before opening the dura, which allowed a more precise approach to the mass. Intraoperative sonography identified the lower end of the syrinx cavities, which allowed optimal catheter placement. Fibroglial scar tissue, which may compartmentalize these syrinx cavities, was clearly shown, and the efficacy of shunt catheter placement was immediately determined. In diastematomyelia, intraoperative sonography identified the relationship of the hemicords to bony, cartilaginous, and/or fibrous septa and demonstrated the effect on the tethered hemicords of (1) removing these septa and (2) constructing a single dural sac from the two dural sacs that had enclosed the hemicords. Since significant surgical decisions are based on these sonographic observations, the authors urge widespread use of intraoperative sonography in patients with spinal dysraphism and syringohydromyelia.

Surgical procedures undertaken to treat dysraphic states may be difficult to perform because several complex anomalies often occur together and because long segments of the spine may be involved. Since the clinical outcome depends on the proper identification, localization, and correction of all such abnormalities, it is desirable to image the spinal canal and its contents *during* surgery. Portable real-time sonography permits the surgeon to monitor the progress of surgery, to extend his procedure as the disease dictates, and to ensure as complete a correction as possible before closure.

In this report, we use *spinal dysraphism* to include all those congenital spine abnormalities in which there is a disorder of midline fusion of mesenchymal, bony, and neural structures [1, 2], such as diastematomyelia, congenital intraspinal masses, tethered cord, and occult sacral meningocele. We review our experience with these abnormalities and with syringohydromyelia in order to demonstrate the great impact intraoperative sonography can have on the surgical management of these disorders.

Subjects and Methods

Twenty-four consecutive patients, with the exception of those with myelomeningoceles, who had surgery for spinal dysraphism or syringohydromyelia were evaluated during surgery with real-time sonography. There were nine patients with spinal cords tethered by lipomas (six), epidermoid tumors (one), dermoids (one), and teratomas (one). Four patients had congenital tumors not associated with cord tethering, including two hamartomas, one dermoid, and one subpial lipoma. Seven patients had syringohydromyelia. Three had diastematomyelia and one had an intrasacral meningocele. Sonography did not prove to be useful in

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evaluating neonates who were undergoing repair of myelomeningoceles, since in these patients the anatomic relationships under inspection are obvious to the surgeon. Therefore, no patients with myelomeningoceles are included in this report.

Real-time sonography was performed with either an ATL Neuro-Sector scanner (19 cases) or an Acuson 128 computed sonography unit (five cases). Techniques for using this equipment during spinal surgery and the sonographic features of the normal spinal cord and spinal canal have been published [3, 4]. Briefly, a laminectomy is performed and sterile saline is then poured into the surgical wound to create a fluid pathway for transmission of sound. A 5.0- or 7.5-MHz transducer is placed into this fluid bath and baseline longitudinal and transverse images are obtained. At appropriate intervals thereafter, the surgical procedure is monitored by sonography and the resultant images are compared with the baseline sonograms to assess the progress of the operation. After closure of the dura, a closing sonogram allows final assessment of the surgical field to rule out a hematoma and to confirm the intended result of the procedure.

Results

Regardless of the type of lesion, the initial intraoperative sonogram demonstrated whether or not the laminectomy was

performed at the proper level, whether the surgical exposure was adequate, and whether the preoperative diagnosis was correct and/or complete.

In patients with tethered spinal cords, sonography showed the relationship of the tethering mass to the low-lying cord (Figs. 1–4); the plane of demarcation between the tumor and normal cord tissue (Figs. 1–3); the presence or absence of gross tumor involvement of the cord (Figs. 1–3); the size, number, and extent of intradural and extradural tumors (Fig. 5); and the results of total or partial tumor removal (Fig. 1). Sonography documented lipomatous involvement of the filum terminale (Fig. 4) and the effect of sectioning the filum.

In cases of congenital tumors not associated with cord tethering (Fig. 6), sonography showed the relationship between the cord and the mass. The echogenicity of the congenital tumors varied. Many of the lipomas were more echogenic than normal spinal cord tissue, but their specific appearances varied. The other solid tumors were also more echogenic than normal spinal cord. One dermoid tumor with calcification exhibited areas of bright reflectivity [3]. No attempt was made to correlate the exact histology of the tumor with specific sonographic characteristics.

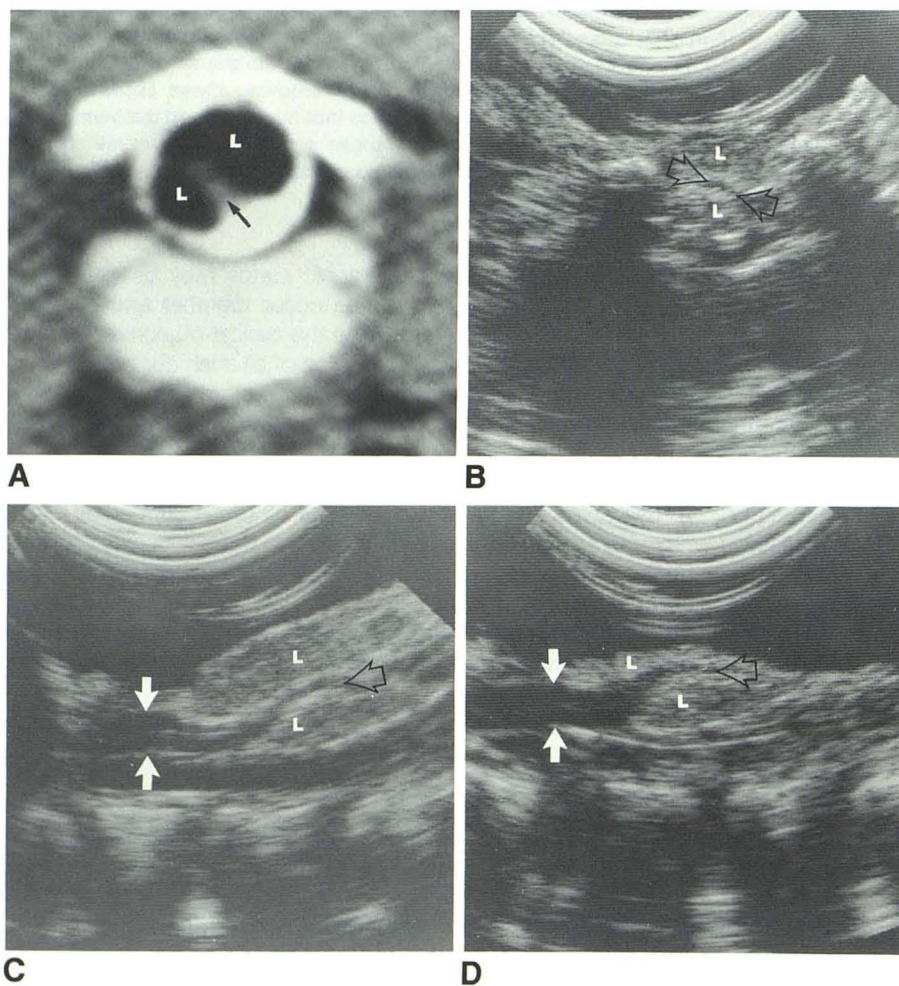


Fig. 1.—Tethered cord with lipoma. Lumbar metrizamide CT scan (A) oriented to conform to transverse sonogram (B) shows lipoma (L) nearly encircling flattened and tethered spinal cord (solid arrow). At surgery, transverse (B) and longitudinal (C) sonograms show lipoma dorsal and ventral to thinned spinal cord (open arrows). After untethering of cord and subtotal removal of dorsal part of lipoma, final sonogram (D) shows residual lipomatous tissue (upper L), no change in ventral lipoma (lower L), and thinned cord (open arrow). In C and D, spinal cord (between solid arrows) cephalad to lipoma appeared normal sonographically. The surgeon decided to resect just the dorsal component of the lipoma for fear that further cord manipulation would induce neurologic deficits.

Fig. 2.—Tethered cord with lipoma. Patient with previous surgery for sacral lipoma.

A, Myelogram shows tethered cord (solid arrows) and intradural mass (open arrows) at level of distal cord.

B, Intraoperative sonogram shows transducer touching large extradural component of lipoma (L), relationship of lipoma to dorsal cord, and plane of demarcation (solid arrows) between lipoma and cord. Tethered cord is identified (between open arrows). Highly echogenic surfaces of cord, its normal echogenicity, and preservation of central echo (ce) indicate that lipoma has not infiltrated cord.

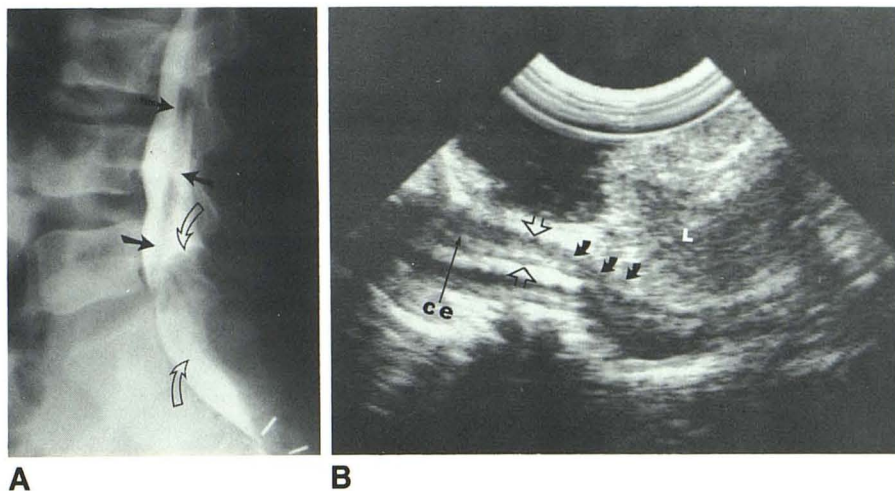


Fig. 3.—Tethered cord with dermoid. Patient with previously repaired myelomeningocele.

A, Myelogram shows tethered cord (black arrows) ending in intradural mass (white arrows).

B, Longitudinal sonogram during surgery shows tethered cord (solid arrows) associated with intrasacral mass (between open arrows) that proved to be dermoid. Note sharp demarcation between distal cord and mass.

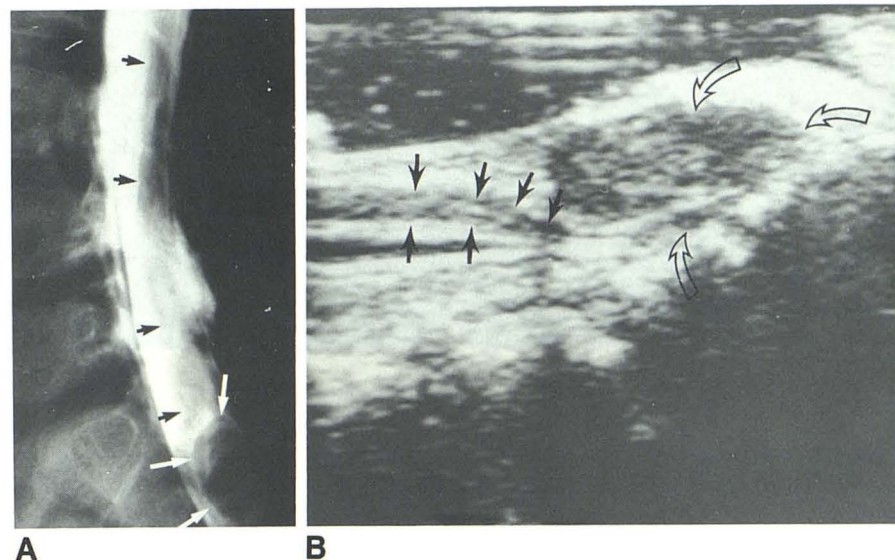
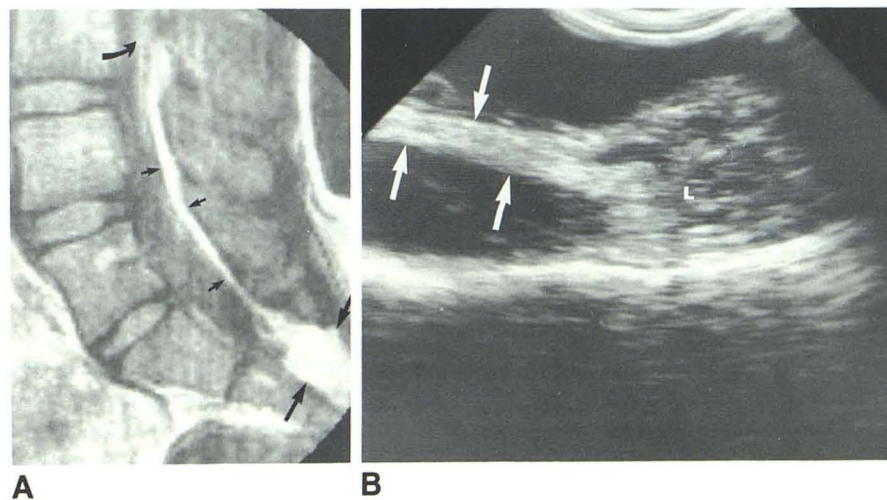


Fig. 4.—Tethered cord with lipoma and lipomatous filum terminale.

A, MR shows lipoma (long straight arrows), lipomatous involvement of filum terminale (short straight arrows), and insertion of filar lipoma into distal tethered spinal cord (curved arrow).

B, Longitudinal intraoperative sonogram shows lipoma (L) to be mass of mixed echogenicity. Distal filum (between arrows) was enlarged by echogenic lipoma. After resecting lipoma, the surgeon transected filum, and rostral portion of filum was seen to move 7–8 mm superiorly.



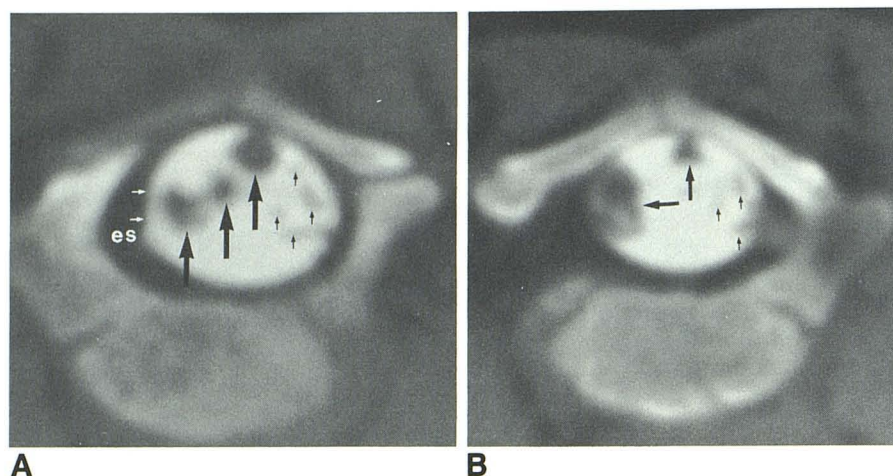


Fig. 5.—Extradural and intradural epidermoid tumors.

A and B, Metrizamide CT shows widened lumbar epidural space (es) on left, with flattening of thecal sac (white arrows). In addition, large intradural masses (large black arrows) were seen and were distinguishable from smaller roots of cauda equina (small black arrows). (A and B are reversed from their normal orientation so they can be easily compared with sonograms.)

C and D, Transverse intraoperative sonograms show flattened thecal sac on left (white arrows) consistent with extradural mass (em) and large intradural masses (black arrows, C). In D, at level shown by tip of curved arrow, extradural mass (em) extended through dura and presented as large intradural mass (im). Dermal sinus was found to extend into extradural space, where it was associated with epidermoid and chain of intradural epidermoid tumors. All tumors were resected.

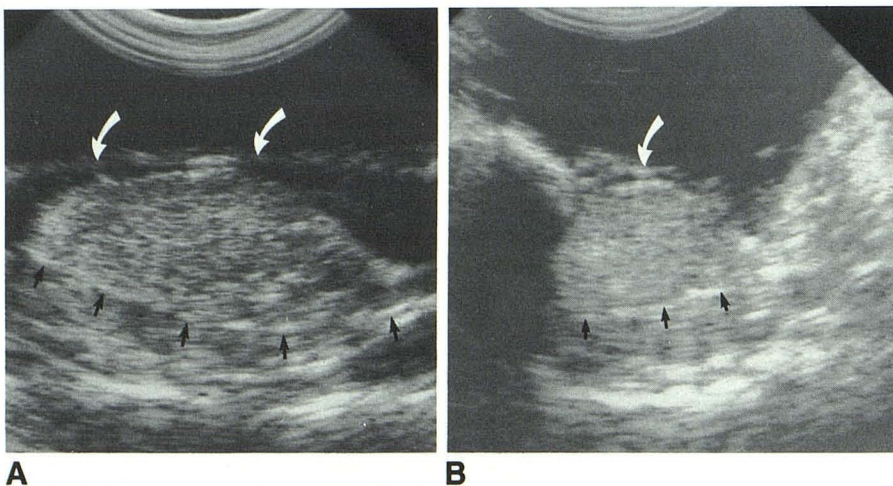
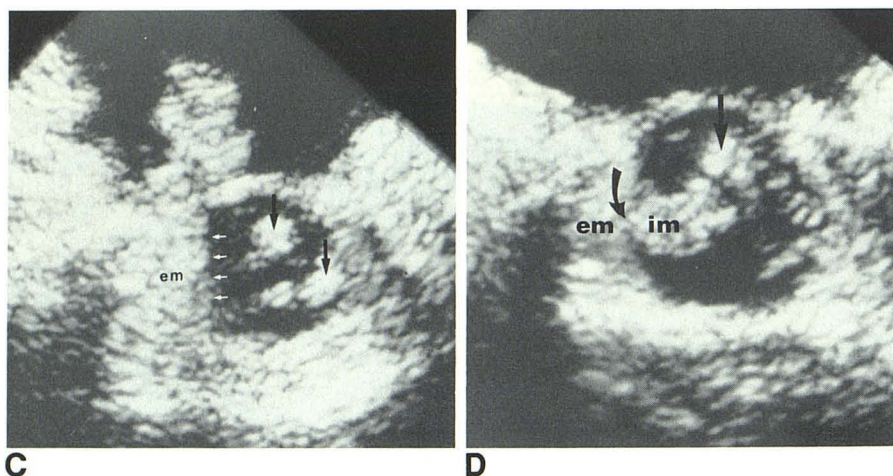


Fig. 6.—Subpial lipoma. Myelography and metrizamide CT (not shown) were diagnostic of intramedullary mass at T12 level. While attenuation coefficient of mass suggested a lipoma, it could not be determined whether lipoma extended deeply into spinal cord or was purely subpial in location. Longitudinal (A) and transverse (B) sonograms show sharp plane of demarcation (black arrows) between lipoma and dorsal portion of cord. There was no evidence of extradural lipoma. Note intact dura (white arrows).

In cases of diastematomyelia (Fig. 7), sonography showed where the hemicords lay within a single dural sac and where they lay within partially or totally separate dural sacs. The extent of the fibrocartilaginous and/or bony septa and the site at which the hemicords rejoined into a single cord were

also identified. The completeness of removal of the septa, adequacy of neural element decompression and/or untethering, and the absence of constriction of the hemicords by the reformed dural sac were confirmed with sonography at the time of closure.

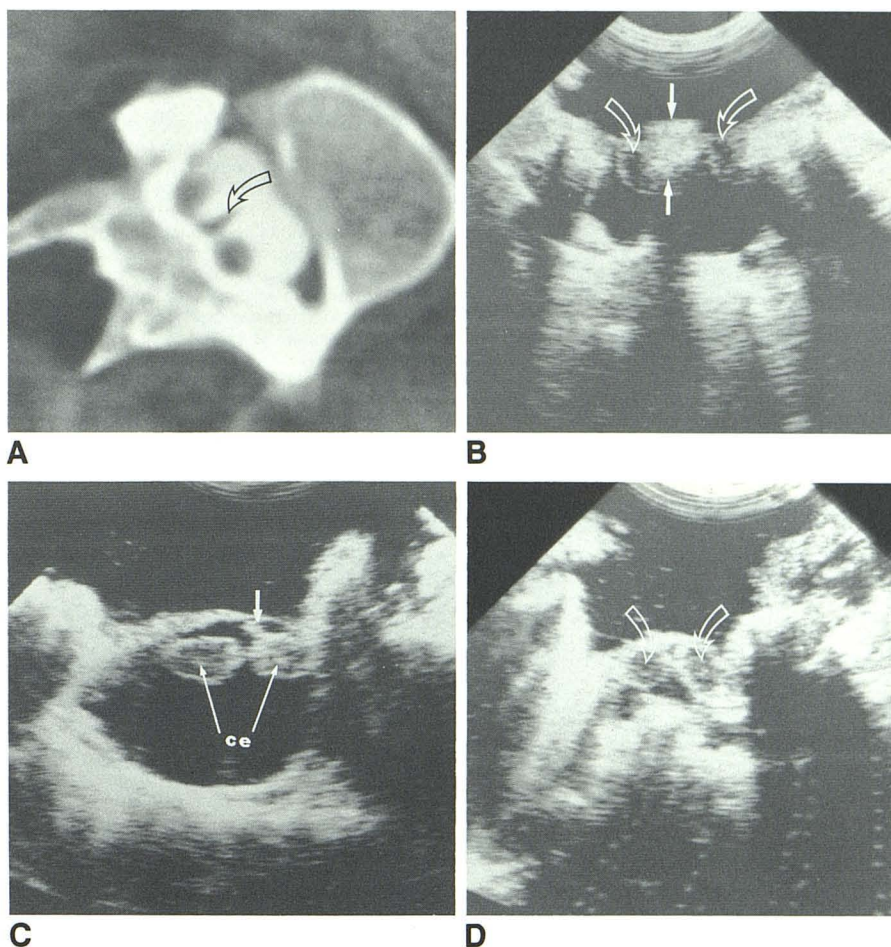
Fig. 7.—Diastematomyelia.

A, Lumbar metrizamide CT showed two hemicords within single dural sac at T12–L1, two separate dural sacs split by large bony septum at L2, and partial separation into two sacs by fibrous septum at L3 (arrow). Distortion of spine was secondary to severe rotatory scoliosis.

B, Intraoperative sonogram shows two hemicords (curved arrows) separated by bony septum (between straight arrows).

C, Thin echogenic structure (short arrow) continuous with dorsal dura-arachnoid layer was believed to present portion of fibrous septum. Note sonographically normal hemicords with their central echoes (ce), hyperechoic surfaces, and hypoechoic interiors. There was no evidence of hydromyelia in either hemicord.

D, After resection of bony septum and formation of single dural sac, the two normal hemicords (arrows) were seen within collapsed thecal sac.



In patients with syringohydromyelia (Figs. 8 and 9), six of the seven cavities were treated with a shunt between the syrinx cavity and the subarachnoid space. In those patients, intraoperative sonography identified the caudal end of the syrinx cavity (midcervical region in one, upper thoracic in three, and conus in three), so the most distal myelotomy possible could be performed. Intraoperative sonography also demonstrated whether a single shunt catheter was adequate for cyst decompression, whether the syrinx was compartmentalized by fibroglial scar tissue, and whether shunting the syrinx relieved the pressure on the tonsils and cervical medullary junction. The use of a single catheter was sufficient to decompress six of our syringohydromyelic cavities. In one patient in whom the syrinx was treated by simple fenestration, sonography demonstrated a decrease in the size of the cyst cavity.

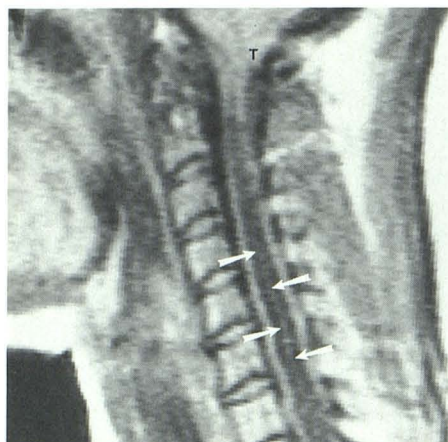
In the single case of intrasacral meningocele (Fig. 10), intraoperative sonography showed the plane of separation between the distal spinal canal and the meningocele and identified the position and course of the sacrococcygeal nerve roots. These roots were best appreciated by their courses and by their pulsatile motion with real-time imaging.

The amount of time the radiologist is required to spend in the operating room performing sonography [5] depends on

the complexity of the abnormality and the skill of the surgeon. While only a portion of his time involves the actual sonographic examination, the total time commitment in these types of cases is at least 1 hr and perhaps as long as 2½ hr.

Discussion

Spinal dysraphism represents a failure of normal midline mesenchymal, bony, and neural fusion. Diastematomyelia, congenital tumors, meningocele, meningomyelocele, and tethered cord are some of the diverse forms of spinal dysraphism. These forms of dysraphism can occur as isolated anomalies or they can occur in combination with other dysraphic states. Thus, a tethered cord may be associated with congenital intradural tumors, with a short, thickened filum terminale that may or may not show evidence of lipomatous involvement, with fibrous bands and adhesions, or with dermal sinuses [1, 2, 6–8]. When a tumor is found, it is usually a lumbosacral lipoma. Less often, a dermoid, epidermoid, teratoma, neuroenteric cyst, or Wilms' tumor [9] may be present. These tumors may act as expanding intraspinal masses, causing compression of adjacent neural tissue, or they may cause traction or tethering of the spinal cord. In patients with dysraphism, surgery is performed primarily to free the spinal cord



A

Fig. 8.—Syringohydromyelia.

A, Preoperative MR shows syringohydromyelia from C2 to T11 level, thin linear structure (arrows) within syrinx cavity, and tonsillar ectopia (T).

B, Before insertion of shunt catheter, longitudinal sonogram shows distal end of syrinx cavity (arrow) at T11.

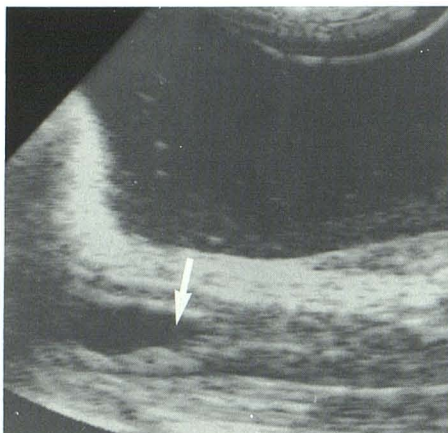
C, Transverse sonogram at C5 shows echogenic structure presumed to be fibroglial scar (between arrows) within syrinx cavity (compare with A).

D, Longitudinal sonogram at cervicomedullary junction shows ectopic tonsils (T) and compression of dorsal cervical cord (arrows). After myelotomy and shunt catheter insertion at T11, the catheter was advanced 25 cm within the syrinx to about the T2 level.

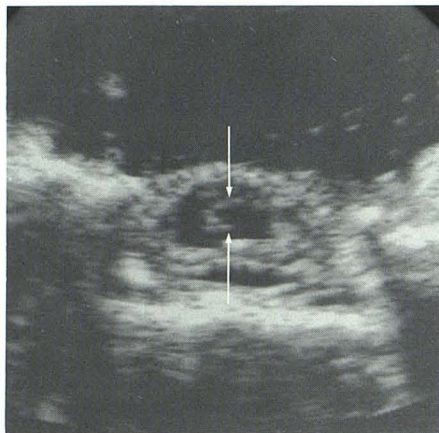
E, Repeat transverse sonogram at T11 shows shunt catheter (arrow) within collapsed syrinx. Collapse of subarachnoid space resulted in poor visualization of cord outline.

F, At C5, collapsed syrinx is seen (compare with C).

G, At cervicomedullary junction there is diminished mass effect on tonsils (T) and spinal cord (arrows) and increased CSF around tonsils and cord (compare with D). Postoperative MR confirmed complete collapse of syrinx.



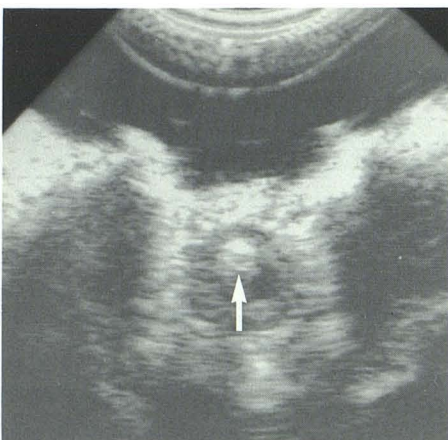
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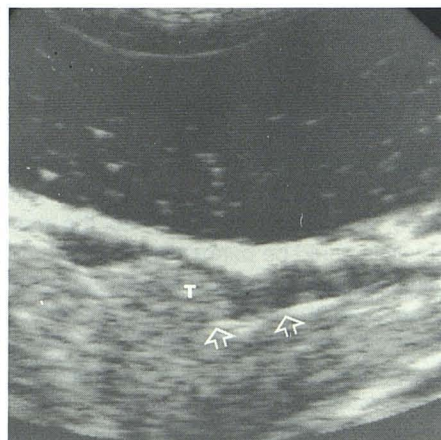
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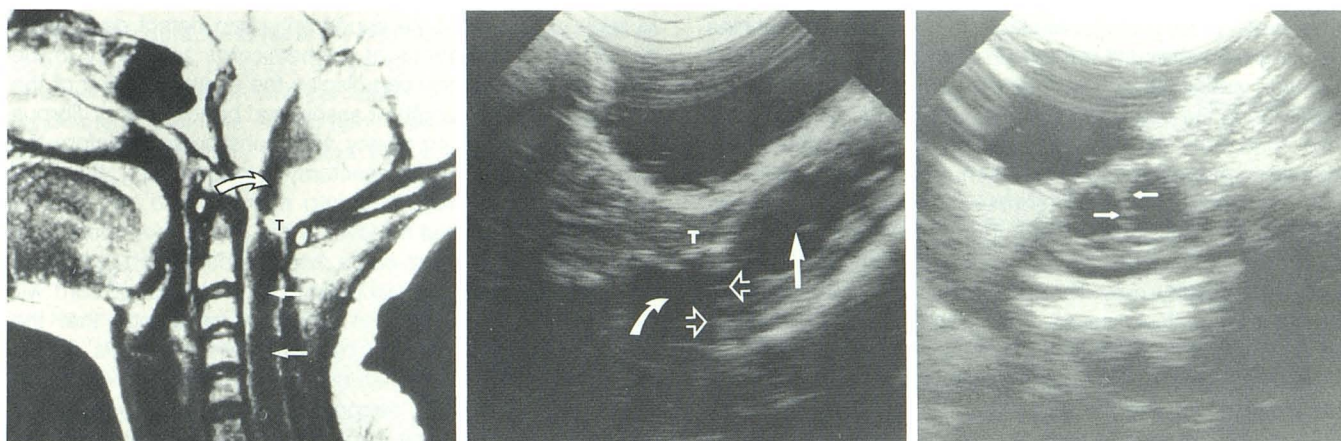
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G

and nerve roots from pathologic tethering or compression. These developmental anomalies cause neurologic deficits that often worsen with age [10–12], and since surgical repair usually arrests progression of deficits rather than restoring neurologic function, surgery should be performed as soon as

the diagnosis can be established. Ideally, this would be in the still asymptomatic patient, before the patient loses motor power or sensation, becomes incontinent of urine or feces, or suffers progressive deformity of the spine or lower extremities [13].



A Fig. 9.—Syringohydromyelia and syringobulbia.

A, MR shows hydromyelia (straight arrows), syringobulbia (curved arrow), and tonsil prolapse (T).

B, After laminectomies at C6 and C1, longitudinal sonogram shows syringohydromyelia (solid straight arrow), syringobulbia (curved arrow), and ectopic tonsils (T). Fibroglial scar (between open arrows) partly separates these two cystic collections in area where MR showed narrowing between syringohydromyelia and syringobulbia.

C, Transverse sonogram at C6 shows fibroglial tissue (arrows) traversing width of syrinx.

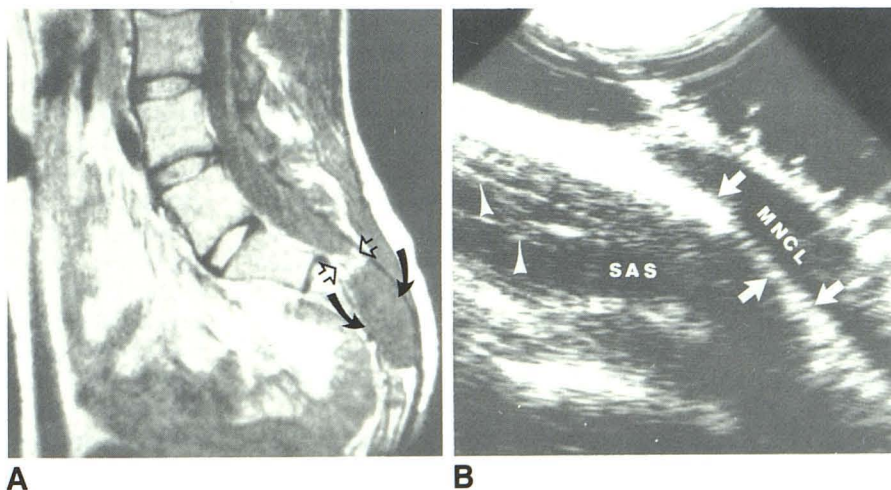
D, After insertion of shunt catheter (solid arrow) into cord at C6 there was complete collapse of left side of syrinx (open arrow) when compared with **C**. Catheter was advanced to C1 level, but sonography showed that, initially, catheter would not pass cephalad to scar seen in **B**. After repeated attempts, the catheter was finally advanced above scar into syringobulbia.

E, Longitudinal sonogram shows catheter (solid arrows) in center of cord with its tip (open arrow) in syringobulbia.

Fig. 10.—Occult sacral meningocele.

A, MR shows CSF-equivalent mass (between solid arrows) within sacral canal, causing erosion of sacrum at S2 and S3. Separating normal ending of subarachnoid space and meningocele is area of high signal intensity within which there are some thin hypointense signals, consistent with fibrous membrane (open arrows). At myelography (not shown) there was a block to the flow of contrast material at S1 with no filling of cyst.

B, Longitudinal intraoperative sonogram localizes soft-tissue membrane (arrows), which separates normal subarachnoid space (SAS) from meningocele (MNCL). Sacral roots (arrowheads) within subarachnoid space were shown not to pass into distal cyst. With real-time sonography, a small opening, acting as a ball valve, was seen to connect subarachnoid space with meningocele. This opening cannot be seen on these static images but was located at about the level of the middle of the three solid arrows. The cyst was totally resected and the small opening was repaired.



Accurate preoperative radiologic studies help in planning the surgical correction of spinal dysraphism [10, 13-16]; however, additional critical management decisions must be made during the operation itself. In such cases the intraoperative sonogram will initially show the surgeon whether an adequate amount of bone has been removed or whether a wider laminectomy will be required to approach the mass successfully. This is particularly important in surgery of the spinal dysraphic states, since the level of bony and/or cutaneous abnormality may be many segments away from the site of the intraspinal disease. Sonography offers the surgeon the opportunity to identify the extent and configuration of associated masses and their relationship to normal neural tissue before opening the dura. This is particularly important when an abnormality is ventral to or out of the surgeon's direct line of vision. In addition, without intraoperative sonography, the safe and effective resection of congenital tumors can be difficult because the margins of the tumors may not be easily distinguishable from normal cord tissue either on gross inspection or with the aid of the operating microscope. With intraoperative sonography, however, these important interfaces are identifiable (Figs. 1-3 and 5) and surgical resection may be accomplished more easily.

Whether or not a congenital tumor is associated with a tethered cord, intraoperative sonography demonstrates whether (1) the cord is surrounded by tumor (Fig. 1), (2) the tumor is related to just one surface of the cord (Figs. 2 and 3), (3) the tumor is in extradural and/or intradural spaces (Fig. 5), and (4) the tumor is subpial in location (Fig. 6). If the spinal cord is surrounded by tumor and the patient is neurologically intact, the surgeon may take a less aggressive approach to tumor removal, particularly if significant retraction of cord tissue would be required to complete tumor removal. Intraoperative sonography will also show whether there is a distinct interface between the tumor and the cord or whether the interface is indistinct, suggesting the possibility of gross infiltration of the cord by the tumor. Gross infiltration precludes total excision and suggests that simple biopsy or a debulking procedure might be more prudent. Since most solid tumors are hyperechoic, intraoperative sonography permits the surgeon to distinguish between tumor and normal cord tissue, to determine the plane of intended tumor resection (Fig. 6), and to determine the site at which to section a lipomatous filum (Fig. 4). In patients with filum lipomas, the exact end of the conus and the beginning of the filum may be indistinct even under the operating microscope. Sonography permits more precise determination of these borders and helps to avoid inadvertent injury to the conus. Unfortunately, our experience to date has shown no specific sonographic features that reliably distinguish one tumor from another. Indeed, even lipomas show varying echogenicity, perhaps because of different amounts and distributions of fibrous and connective tissue within them or differing dispersions of fat and aqueous elements [17, 18].

In patients with diastematomyelia, the hemicords may be tethered by (1) adhesions to either a bone spur, a fibrocartilaginous spur, or a fibrous septum; (2) a thickened filum terminale; and/or (3) associated congenital tumors [1, 8, 9].

In such cases, real-time sonography can detect dampening of the normal pulsations of the hemicords secondary to the tethering. Sonography can display the sagittal cleaving of the cord, the site and length of associated bone spurs or fibrous septa, and the nature of any adhesions present, before the dura is opened. If the laminectomy is long enough, the point of reunion of the hemicords into a single cord can also be seen. After successful removal of the septum (or spur), lysis of adhesions to the hemicords, and reconstruction of a single dural sac (Fig. 7D), normal pulsations of the spinal cord will be observed. This provides additional assurance that the untethering procedure has been successful. In this series, one patient with diastematomyelia showed anechoic areas within both hemicords, suggestive of an associated hydromyelia. Recent evidence suggests that hydromyelia may occur in 50% of patients with diastematomyelia [19]. Depending on the size of the syrinx, the surgeon may opt to shunt or fenestrate the intramedullary cavity with a microlaser at the same time he untethers the hemicords. However, if intraoperative sonography shows the cyst is small, the surgeon may elect to leave it untreated, since the risk of treatment may then be greater than the potential benefit. It is hoped the additional information provided by sonography will help to prevent late clinical deterioration caused by incomplete removal of the septum and its dural sleeve or inadequate treatment of the other abnormalities associated with diastematomyelia.

Over the years, surgical procedures used to decompress syringohydromyelic cavities have included posterior fossa decompression, terminal ventriculostomy, cyst fenestration, and shunting of the syrinx into the subarachnoid space via a catheter. In our institution, we use sonography to identify the distal end of the syrinx cavity, so a shunt can be passed into the cavity through the lowest possible portion of the spinal cord, minimizing the risk of neurologic deficit (Fig. 9B). Sonography is also used to identify any scars (Figs. 8C and 9B) that may compartmentalize the syrinx into separate or intercommunicating cavities and to determine whether the catheter has adequately decompressed the syrinx (Figs. 8E, 8F, 9D, and 9E). If the single catheter has not proved to be sufficient, sonography is used to monitor the success of further catheter maneuvers (Fig. 9) or insertion of a second catheter. With sonographic guidance, a single syrinx to subarachnoid space shunt catheter successfully decompressed six of the seven syringohydromyelic cavities. In one case, the syrinx was fenestrated and the successful decompression of the cyst was confirmed by sonography before closing.

In one patient with an intrasacral meningocele, sonography helped the surgeon separate the cyst from the distal portion of the thecal sac and to identify the point of connection between the subarachnoid space and the cyst (Fig. 10). Real-time sonography demonstrates the pulsatile motion of nerve roots within the subarachnoid space and/or the cyst, so their position and course can be determined before undertaking cyst resection and repair of the fistulous opening.

In conclusion, we have found that in patients with congenital anomalies of the spine, intraoperative sonography permits the surgeon to see the spinal canal and its contents before

opening the dura, determine the best approach to the pathology, monitor the progress of the operation, and determine whether the operation was partially or wholly successful. We recommend real-time sonography as an "operative fluoroscope" during surgery for spinal dysraphism and syringohydromyelia.

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