CE 2.5 HOURS Continuing Education

Part 3: Examination of the Newborn With Closed Spinal Dysraphism

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ABSTRACT

Spinal dysraphism, an incomplete closure of the neural tube, can be open, exposing the neural elements to the environment, or can be closed, covered with skin. Abnormal development of the spine occurs early in life and also interferes with usual development of the skin. This often creates cutaneous markers or stigmata over the area. Cutaneous markers may include a subcutaneous mass, abnormal hair growth, skin dimple, tag or sinus, or unusual pigmentation. Recognizing these markers is important because, although many closed spinal dysraphisms are asymptomatic at birth, neurological sequelae can occur. The sequelae are insidious and often permanent.

This article, Part 3 in a series of articles devoted to spinal assessment, reviews closed spinal dysraphisms. The article emphasizes identification of various cutaneous markers associated with closed spinal dysraphisms. Early detection and follow-up may prevent neurologic sequelae for the infant later in life.

KEY WORDS: cutaneous marker, cutaneous stigmata, dermal sinus, dysraphism, infant, lipoma, neonate, newborn, physical examination, sacral dimple, split-cord malformation, tethered cord

Care of the newborn places the nurse in a unique position to recognize signs of closed spinal dysraphism (CSD). Although CSDs are often asymptomatic at birth, cutaneous markers such as subcutaneous masses, abnormal hair growth, skin dimples, tags and sinuses, or unusual pigmentation may be present.[1,2] The incidence of closed spinal dysraphism is unknown but is estimated to be 0.05 to 5 per 1000 births.[2-4] Underlying pathology can be related to spinal cord or vertebral abnormalities, cysts, tumors, or a tract between the skin and central nervous system that predisposes the infant to meningitis. These lesions may pose neurological or infectious risks later in life. Once neurologic deficits occur, they may be irreversible. Early detection and management may prevent sequelae such as tethered cord or neurogenic bladder later in life.

Discrepancies exist between dermatological and neurosurgical literature regarding the significance of cutaneous lesions. These lesions overlay the spinal column in 50% to 100% of infants with closed spinal dysraphism.[2,3,5] Experiential differences exist because of referral of patients with significant dermal lesions to dermatologists and referral of those with neurological symptoms to neurosurgeons. Lack of consistent nomenclature hinders consensus between the 2 specialties.[6,7] Table 1 lists terminology used to describe CSD.

It is important that the neonatal nurse be aware of the array of markers, particularly those that have a correlation with an underlying anomaly of the spine. Close examination and detection of cutaneous markers and a precise description and effective communication with the medical team is essential in early detection of closed spinal dysraphisms. Early detection and treatment can reduce morbidity and avoid injury related to infection, cord compression, and tethered cord. If occult spinal dysraphism is present, neurological and neurosurgical follow-up is necessary to monitor for spinal cord dysfunction, especially motor and sphincter control.[7]

This article is the third in a series on the newborn spine. Part 1 reviewed spinal cord injury,[8] and Part 2 reviewed open spinal dysraphism.[9] This article reviews the embryology of closed spinal dysraphisms and the significance and clinical implications of physical findings. The significance of an abnormal position of the conus medullaris and a tethered cord also are discussed.

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From Texas Children's Hospital, Houston, Tex.

TABLE 1. Terminology Associated With Closed Spinal Dysraphism[1,2,4,5,7,10,14,15,27]

Term	Synonym	Description
Closed spinal dysraphism	Occult spinal dysraphism	Congenital malformation of spine arising from incomplete development of dorsal midline structures; covered with skin
Conus medullaris	Medullary cone	Tip of the spinal cord
Cutaneous marker	Cutaneous lesion Cutaneous stigmata	Abnormal pigmentation, skin characteristics and hair distribution
Dermal sinus tract		Opening from spinal canal to skin; associated with recurrent meningitis
Filum terminale		Fibrous thread indicating the line of regression of the caudal end of the spinal cord extending from the conus medullaris and attaching to the periosteum of the first coccygeal vertebra
Hamartoma		Tumor that does not usually contain all 3 germ-cell layers; tissues are well differentiated and may grow, but malignancy is rare
Lipoma	Lipomeningocele Lipomyeloschisis	Benign, encapsulated tumor of adipose tissue
Myelocystocele		Localized cystic dilation of the central canal of the spinal cord
Neuroenteric cyst		Incomplete separation of endodermal elements of the notochordal plate; the lesion is lined with endodermal epithelium
Spina bifida occulta		Incomplete vertebral opening covered with skin
Syringocele	Syringomyelocele Syrinx	A fluid-filled cavity within the spinal cord; terminal syringocele occurs in the distal third of the spinal cord
Teratoma		A potentially malignant lesion that contains tissue from all 3 germ layers
Tethered cord	Tethered cord syndrome Occult spinal dysraphism sequence	Movement of the filum is restricted or tethered by fibrous bands or a mass such as a teratoma or lipoma
Diastematomyelia	Type I split-cord malformation	Type I, each hemicord is contained within its own dural sac and separated by a bony or fibrocartilaginous spur
Diastomyelia	Diplomyelia Type II split-cord malformation	Both hemicords are contained within a single dural sheath; does not necessarily have a spur or spur may be fibrous

EMBRYOLOGY OF SPINAL DYSRAPHISMS

Elements of the spine, cord, and skin originate in common embryonic cells.[6,7] When the neural tube is closed, superficial ectoderm separates from neuroectoderm, fusing in the midline to form the skin covering the neural tube. A disruption in this process occurs when the neural tube does not close normally, interfering with normal development of the overlying skin and creating cutaneous markers in the area of malformation.[6]

During embryogenesis, a number of intricate processes take place during the formation of the spine. Interruption of these processes may interfere with normal development of the spine and related structures. Of particular importance is the potential for remnants of adhesion between the notochord and the endoderm to persist during the second week of gestation. These remnants may create a spur that divides the developing cord into 2 parts. This is known as *split-cord malformation* and is associated with longer spinal cord segments, lower conus medullaris location, and an increased rate of malformations in the lower spine[10] (Figure 1).

During secondary neurulation, which occurs from day 25 to day 48,[9,11] vacuoles, which have formed within the tail bud, cannulize. The canal then connects the tail bud to the neural tube that formed earlier during the process of primary neurulation. The caudal region of the canal (below S2) then forms through a process called regression. The conus medullaris, terminal filum, and caudal equina are formed during this process.[4,10,11]

FIGURE 1.

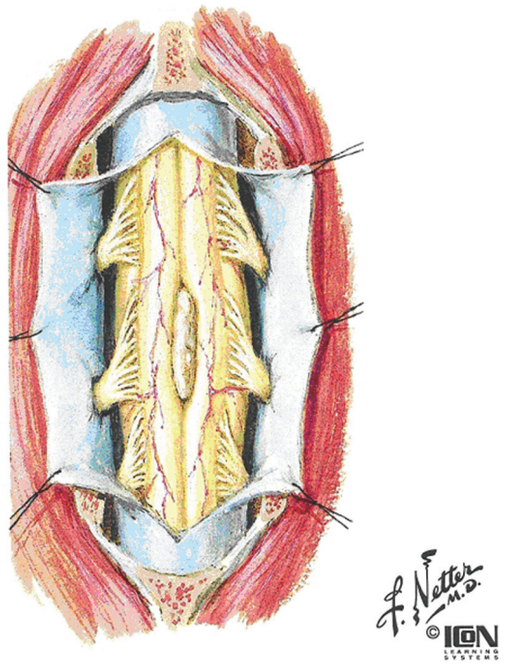


Diagram of split cord. Note the spur dividing the cord into 2 hemicords. Reprinted with permission from: Netter, FH. Primary neurological disorders in infancy and childhood. In: *The Netter Collection of Medical Illustrations—Nervous Systems and Neurological Disorders.* Havas MediaMedia, Icon Learning Systems. Copyright 1986.

FIGURE 2.

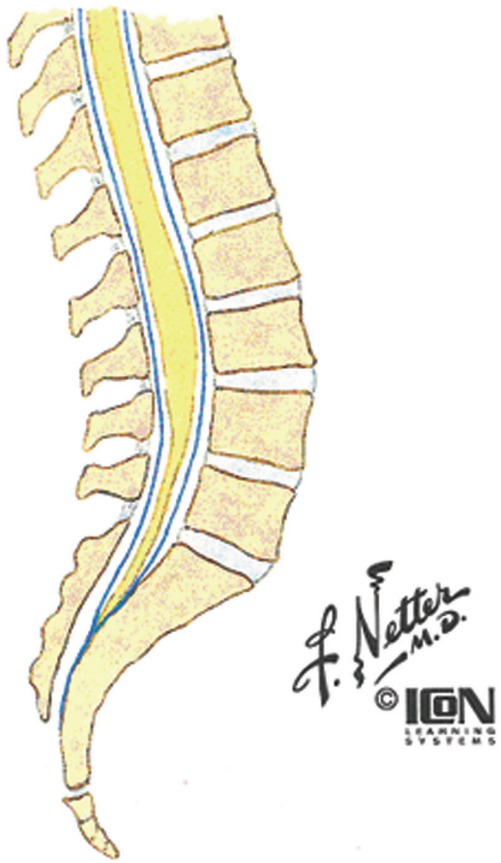


Diagram showing tethered cord. Tethering prevents normal ascension of the conus medullaris. Reprinted with permission from: Netter, FH. Primary neurological disorders in infancy and childhood. In: *The Netter Collection of Medical Illustrations—Nervous Systems and Neurological Disorders.* Havas MediaMedia, Icon Learning Systems. Copyright 1986.

Maldevelopment may result in CSD and is frequently associated with a tethered spinal cord (Figure 2). Tethering of the cord is caused by tissue attachments that limit ascent of the spinal cord within the spinal column. Inelastic structures, such as a fatty filum terminale, tumors, myelomeningoceles, lipomyelomeningoceles, scar formations, or a bony septum, anchors the lower segment of the spinal cord.[12,13] The bony spine grows faster than the cord, and this attachment or tethering results in traction on the spinal cord as the infant grows. This traction can impair circulation, causing progressive cord ischemia and neural dysfunction.[1]

Spectrum of Spinal Dysraphisms

The term *spinal dysraphism* is a general term used to describe incomplete closure of the spinal axis and may encompass a number of malformations of the spine.[14] Tethering of the cord is a common feature of spinal dysraphisms.

Split-Cord Malformation

Split-cord malformation ranges from a simple cleft in the spinal cord to complete duplication of the spinal cord into 2 hemicords. Tripomyelia (triplication) is also possible but rare. A bony or cartilaginous spur separates the hemicords. The cords may be in a single dural sac or each may have its own dural sheath. Abnormalities of ventral and dorsal nerve roots may occur. The cords may be symmetric or asymmetric in size. Split-cord malformation may be isolated or associated with other dysraphisms, scoliosis and tethered cord (75%), or low-lying spinal cord. Midline cutaneous markers occur in 90% of infants with split-cord

malformation, with the marker occurring at the level of bifurcation.[15-17]

Lipoma

Lipoma, the most common form of closed spinal dysraphism, is a benign, soft, rubbery tumor composed of mature fat cells that are thought to result from embryonic stem cells. A lipoma may lie in the dermis or in the spinal canal, where it can cause cord compression and tethered cord.[4,14,18] Familial cases have been reported, and incidence in these cases is not reduced by folate supplementation. A lipoma may overlay a myelomeningocele.

Neuroenteric Cysts

Neuroenteric cysts are rare congenital lesions of the spine that are lined with endodermal epithelium. They are usually located in the thoracic or cervical regions, and most are ventrally located. The cysts derive from endodermal inclusions within the spinal column. Neuroenteric cysts may result from a persistent communication between endodermal and neuroectodermal tissues and be associated with other congenital spinal anomalies. They are more common in males and may present as a mediastinal mass with a concurrent vertebral anomaly. Deviation of an oral gastric tube may signal the presence of a mediastinal mass.[19-21]

Teratoma

Teratomas, the most common tumor in newborns, vary in size, location, and tissues involved.[5] Eighty percent occur in females.[22] A full discussion of this topic is beyond the scope of this article (Figure 3).

Dermal Sinus Tracts

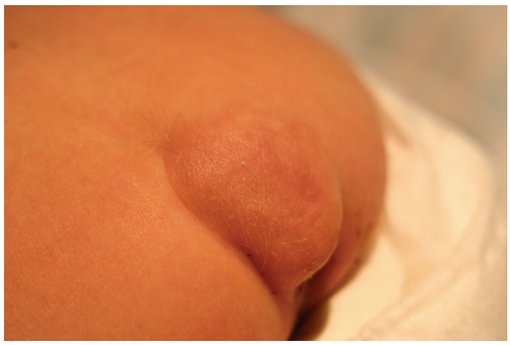
Dermal sinus tracts usually appear as a midline dimple, with or without a tuft of hair, and may have pigmentation of the surrounding skin. Findings include cutaneous markers, leaking of cerebral spinal fluid (CSF), and neurologic deficit or infection.[4,7,23]

Although most dermal sinus tracts are in the lumbar or lumbar-sacral area, they can occur at any location on the spine. They are generally above the gluteal cleft and have a cephalic orientation. Over 50% extend into the spinal canal, placing the infant at risk for meningitis.[4] Clinically these are suspected based on the presence of overlying cutaneous markers or when signs of local infection such as redness or induration occur. Significant neurologic sequelae may develop as a result of recurrent meningitis if diagnosis and treatment are delayed.[4,7,23]

Myelocystoceles

A myelocystocele is a localized cystic dilation of the central canal of the spinal cord. Terminal myelocystoceles are located in the lumbosacral area. Nonterminal myelocystoceles are located in the thoracic area and are rare.

FIGURE 3.



Sacral teratoma. Note variations in skin color. Sacral teratomas are potentially malignant. Photo courtesy of Dr. David Clark. Used with permission.

Risk Factors

Risk factors for CSD are similar to those for open spinal dysraphism and are explored in detail in Part 2 of this series on the spine. These include a familial history of neural tube defects, folic acid or zinc deficiency in the mother, use of anticonvulsant medication by the mother, and maternal diabetes. Genetic factors under investigation include those that affect folic acid metabolism and methyltransferase reactions involving methionine and homocysteine.[2,9]

Prenatal Testing

Because skin covers these anomalies, alpha fetal protein and acetylcholinesterase levels are not elevated. Some closed spinal dysraphisms may be identified when prenatal ultrasound is performed at about 20 weeks gestation.[24]

Examination of Infants with Closed Spinal Dysraphism

During a newborn physical exam, the presence of cutaneous markers overlying or adjacent to the spine may be the only clue to the presence of a CSD.[25] Lesions are generally midline and may be subtle in appearance. It is important for the nurse to distinguish normal variations of the skin from those that are abnormal and to recognize cutaneous markers that are likely to be associated with underlying pathology.[6,14]

Perform a focused physical examination of the spine. Roll the infant onto the abdomen if the condition allows. Palpate each vertebrae (Figure 4) and note any protuberant or absent processes.[2] Visually examine the spine, looking for any midline dermal lesions.[25]

FIGURE 4.

Palpate each spinal process to assess for spina bifida occulta. Photo courtesy of Carol Carrier. Used with permission.

While most of these occur in the lumbosacral area, they may occur any place along the spine.[14] Document the presence of any skin variations in the medical chart using objective terms to describe size, location, shape, mobility, texture, and color. Cutaneous markers can vary. Specifically examine for:

- unusual pigmentation
- abnormal hair distribution or texture
- alterations in skin integrity
- variations in skin surface characteristics, such as folds, pits or dimples, skin tags or appendages
- deviation or asymmetry in the gluteal fold

Table 2 describes overlying cutaneous markers and their synonyms.

Observe for variations in skin color, which can range from pale to purple. Note the size of the lesion and whether it crosses the midline of the spine. Palpate the margins to see if the area is raised or flat. Pigmented lesions can be described as patches when greater than 1 cm in diameter or nevus when less than 1 cm in diameter.[2] Note any variations in skin color that may occur. Hyperpigmented macules (Mongolian spots) are bluish skin discolorations commonly found on the buttocks of dark skinned infants. These are not indicative of underlying pathology. Nevus simplex, often called stork's bite, may occur on the nape of the neck and is usually benign as well (Figure 5). Both hyperpigmented and hypopigmented areas may occur in conjunction with CSD, and both may be present at the same time. Hyperpigmented lesions include a hemangioma (Figure 6), a port wine stain, or telangiectasis. A hemangioma has a greater correlation with CSD than a flat midline capillary malformation, such as a port wine stain. A nevus anemicus is an example of a hypopigmented cutaneous marker. It is an uncommon cutaneous marker consisting of circumscribed, pale macules with irregular margins sometimes surrounded by satellite macules. Although vascular structures are normal, the vessels are sensitive to catecholamines and remain constricted, producing a pale color. Applying heat or cold or rubbing the lesion causes erythema in surrounding tissues but not in the lesion itself. By itself, nevus anemicus is benign, although there may be an association with neurofibromatosis. In combination with telangiectatic nevus, nevus anemicus may indicate a disruption in normal skin development.[26]

Observe for abnormal hair distribution or texture. Hypertrichosis, an excess of hair or a hairy patch, is a common feature of cutaneous markers associated with CSD (Figure 7A-B). A "faun's tail" or faun's tail nevus, a patch of particularly thick hair, may be dark or light. Note the texture of the hair, it is usually silky.[14] Although cutaneous markers are not specific to any lesion, a faun's tail is thought to be associated with split-cord malformation.[7]

Examine for skin tags or tail-like structures. An acrochordon is a soft pedunculated skin tag and is sometimes categorized as a human tail or neuroectodermal appendage[7] (Figure 8). Most of these lesions are comprised of epidermis and a dermal stalk. A true human tail is rare and is comprised of muscle and nerve fibers with a fatty core.[14]

Inspect for cutis aplasia congenital, a disruption in skin coverage, which may be found in the lumbar region. It is often covered by a thin transparent membrane or looks like a scar. Although rare, there is a high correlation with occult spinal dysraphisms.[7,14]

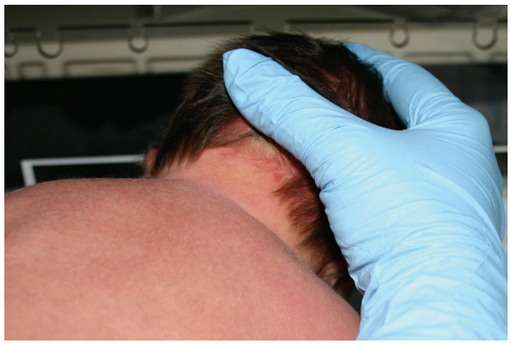
Note the presence of a sacral dimple; a localized pitting in the skin.[7] Document whether the dimple is midline and note the relative size and location of the dimple to the anus. A midline dimple is a common finding. Shallow dimples may occur in 4.3% of normal infants.[8] A small or simple dimple usually occurs just above the gluteal furrow and is less than 5 mm in diameter and 2.5 cm or closer to the anus. A closed spinal dysraphism is unlikely when the dimple is small, shallow, ends in a blind pouch, and is low on the spine.[27] Deep dimples, or atypical dimples, are larger than 0.5 cm and may actually be dermal sinuses[14] (Figure 9). Evaluate dimples or pits for depth and integrity. Avoid probing the pit, which might introduce bacteria.[14] Note any hair protruding from the indentation. Watch carefully for any fluid leakage. A consistently wet area high on the diaper may suggest CSF leakage from a dermal sinus. Dermal sinus tracts are risk factors for recurrent meningitis. If a dermal sinus is suspected, monitor for clinical signs of meningitis, which include fever, vomiting, high-pitched cry, seizures, and poor feeding.

Observe for symmetry of the gluteal cleft. Evaluate the gluteal cleft for bulges or lateral displacement. (Figure 10). Curving of the gluteal cleft suggests a tethered cord or an underlying mass such as a lipoma.[7,14,17] Examine the gluteal cleft carefully for skin tags or sinuses because these may be hidden in the crease

TABLE 2. Cutaneous Markers Associated With Closed Spinal Dysraphism[1,2,4,5,7,10,14,15,16,26,27]

Lesion	Level of Suspicion	Synonym	Description	Association
Curved gluteal cleft	High	Deviation of gluteal fold	Gluteal cleft is distorted by tethering of the spinal cord or an underlying structure.	Spinal lipoma
Cutis aplasia congenita	High	Epitheliogenesis imperfecta Skin dysplasia	Localized failure of skin to form. The area is often covered by a thin translucent membrane, scar tissue, or granuloma.	
Dermal sinus	High		Epithelial-lined dermal tract from the skin to the spine.	Dermoid tumor Pathologic filum terminale
Hemangioma	High		Pigmented lesion found in 50% of patients. Most in association with hypertrichosis	
Human tail	High	Pseudo tail True tail Neuroectodermal appendage Caudal appendage	Term "true tail" is used for cases associated with spinal dysraphism and "pseudo tail" is used when there is no intraspinal connection.	Tethered cord
Hypertrichosis	High	Hairy patch Faun's tail	Localized tuft of hair, common cutaneous lesion suggesting underlying dysraphism.	Split-cord malformation Pathologic filum terminale Dermal sinus
Large sacral dimple	High	Atypical dimple Pilonidal dimple Coccygeal pit	Location: >2.5 cm from anus Size: >5 cm in diameter Atypical dimples are usually above the gluteal crease.	
Lipoma	High		Localized subcutaneous fat tissue presenting as soft mass	Spinal lipoma
Nevus anemicus			Circumscribed, pale macule with irregular margins	
Nevus simplex	Low	Salmon patches Stork bite Nevus flammeus Telangiectasis	Permanent dilation of small blood vessels	
Pigmentary nevus	Low	Birthmark Melanocytic nevi	Nevus containing melanin	
Port wine nevus	Low	Port wine stains Vascular nevus	Benign irregularly shaped macular vascular lesion with smooth surface; blanches with pressure	Spinal lipoma
Skin tag	High	Acrochordon	Small, generally benign growth may be a soft pedunculated skin tag	Spinal lipoma
Small sacral dimple	Low	Coccygeal pit Simple dimple	<2.5 cm from anus; usually within the gluteal crease	
Telangiectasia	Low	Salmon patches Stork bite Nevus flammeus Nevus simplex	Permanent dilation of small blood vessels	

FIGURE 5.



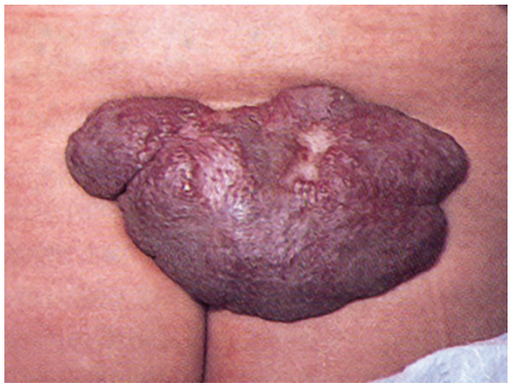
Lift hair from back of neck to assess for cutaneous markers. Nevus simplex as pictured here is generally benign. Photo courtesy of Carol Carrier. Used with permission.

(Figure 11). A deep or twisted gluteal fold also may result from a mass. Examine the genitalia and rectum because there may be associated anomalies of the genitourinary system.[2,14]

Palpate the base of the spine for areas of adipose tissue. Congenital lipomas may be poorly circumscribed and feel like an area overlying a subcutaneous fat pad[14] (Figure 12).

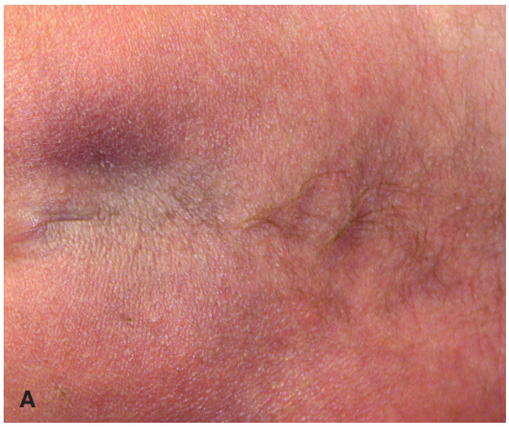
Look carefully for neurological and orthopedic symptoms because they may be subtle. Evaluate the tone of the lower extremities. Observe for symmetrical movement. Note any weakness in the lower extremities because lower extremity weakness or

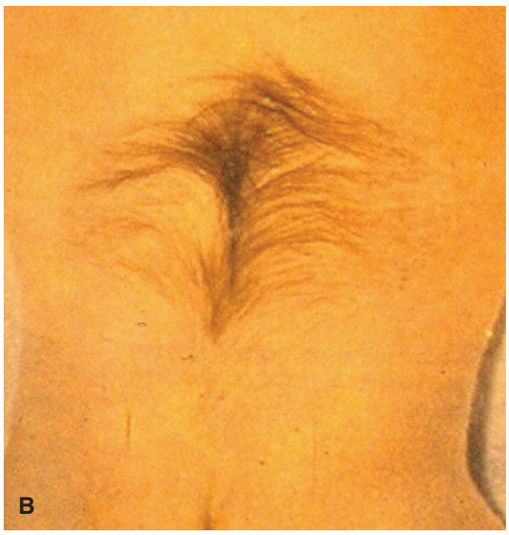
FIGURE 6.



Hemangioma overlying a lipomeningocele. Reprinted from Drolet, copyright 2001, with permission from Elsevier.

FIGURE 7.



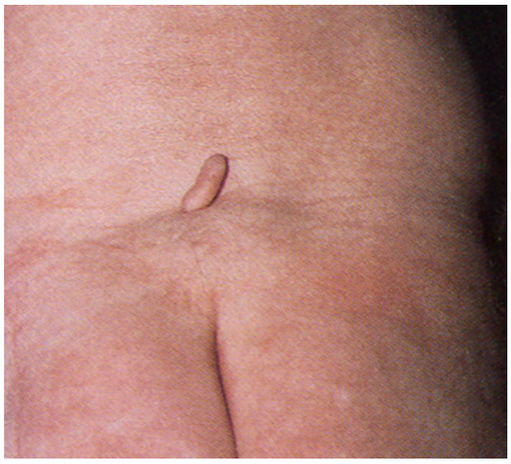


(A) Note the presence of excessive lumbar hair. This may occur alone or in conjunction with another cutaneous marker. Photo courtesy of Dr. David Clark. Used with permission. (B) An excessive amount of hair on the back is known as hypertrichosis. Photo from Baylor College of Medicine's Perinatal Outreach Program's collection. Used with permission.

asymmetrical tone or movements may be seen in infants with spinal dysraphism. Carefully document a baseline examination for comparison with results of later examinations. Asymmetric tone may be associated with abnormal neurulation of 1 hemicord in split-cord malformation. Changes in tone over time may occur with tethered cord syndrome.

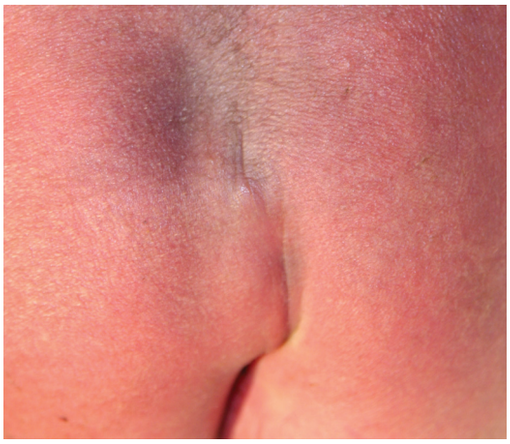
Review the clinical findings and notify the medical team. Two or more lesions or a single lesion with any neurological symptoms has a high correlation to CSD.[7]

FIGURE 8.



Skin tags and appendages may be found in the lumbar sacral area and vary histologically. These are sometimes called *human tails.* Reprinted from Drolet, copyright 2001, with permission from Elsevier.

FIGURE 10.



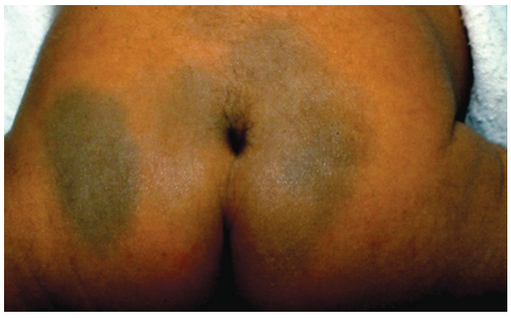
Curved gluteal cleft suggests underlying lesion, such as a lipoma. Photo courtesy of Dr. David Clark. Used with permission.

DIAGNOSIS

Although most physicians agree that the presence of 2 or more cutaneous markers warrants diagnostic follow-up,[7] many argue that using ultrasound is a reasonable screening strategy in infants with single cutaneous markers. As many as 8% of single cutaneous markers are associated with closed spinal dysraphism.[27] The following lesions have higher correlation with CSD, indicating the need for diagnostic screening.[1,14]

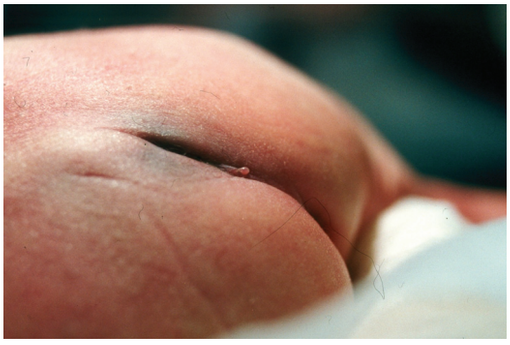
- Sacral dimple more than 2.5 cm from the anus
- Hemangiomas
- Cutis aplasia
- Raised lesions, such as subcutaneous masses, skin tags or tails
- Hairy patches
- Multiple cutaneous markers

FIGURE 9.



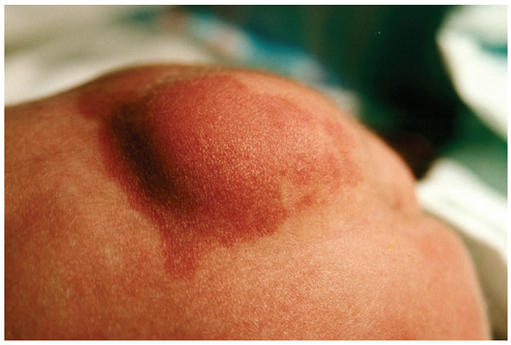
Sacral dimple. Examine carefully for tufts of hair or leaking spinal fluid but do not probe because bacteria may be introduced. The photo also shows Mongolian spots. It is important to document the presence of these areas because someone unfamiliar with Mongolian spots may mistake them for bruises and raise concerns of child abuse. Photo from Baylor College of Medicine's Perinatal Outreach Program's collection. Used with permission.

FIGURE 11.



Carefully examine the gluteal cleft. Skin tags, as shown here, or dermal sinuses may be missed on cursory examination. Photograph courtesy of Dr. David Clark. Used with permission.

FIGURE 12.



Telangiectasis may occur in conjunction with other cutaneous markers. In this picture it can be seen overlying a subcutaneous mass. Diagnostic imaging is needed to identify the underlying structure and a neurosurgical consult requested. Photo courtesy of Dr. David Clark. Used with permission.

Ultrasound is a low-cost and noninvasive method of screening for CSD. Screening may identify the need for additional diagnostic testing in infants who are at risk for permanent neurological deficits associated with CSD.[2,6,27]

Diagnostic Imaging

Although a horizontal sacrum on lateral radiographs may suggest a tethered cord, x-rays are generally not helpful because of the lack of ossification of the posterior spine.[2,17] Computed tomography (CT) scanning is useful for the evaluation of vertebral bodies, posterior arch abnormalities, and spur characteristics in split-cord malformations.

Ultrasonography (US) and magnetic resonance imaging (MRI) are the chosen examinations for the screening and diagnostic confirmation of CSD.[17,27] Spinal US provides dynamic evaluation of lower spinal cord mobility. Both 2- and 3-dimensional US are effective in the evaluation of congenital disorders of the spinal cord. Superior visualization of vertebral bodies and posterior spinal elements can be obtained with 3-dimensional US. The advantages of US include the cost and portability and that no sedation is required. US is a screening tool. If an abnormality such as a low lying or blunt conus medullaris, thickened or fatty filum terminale, lipoma, or lack of pulsatile movement of spinal fluid is found, MRI should be performed.[2,27-29]

An MRI is the most sensitive diagnostic test and is considered the gold standard.[1] Advantages are that MRI is noninvasive and nonirradiating. MRI gives information about the anatomy of the spinal cord, dural sac, conus medullaris and filum terminale.[7,16,18,28-32]

Outcome and Follow-Up

Patients with CSD require lifelong evaluation for the onset of neurological signs.

Recurrent meningitis may occur if a dermal sinus tract is present. Cognitive development may be affected by meningitis. Early surgical intervention is relatively low risk and can prevent the development of permanent deficits.[29]

Most neurological symptoms associated with CSD result from the presence of a tethered cord. Progressive stretching of the lower spinal cord occurs during growth in height, resulting in back pain, sensory loss in the lower extremities, abnormal gait, foot drop, limb atrophy, spasticity, scoliosis, or bowel or bladder incontinence.[1,7,10,29] Refer radiological findings of a tethered cord to a pediatric neurosurgeon for ongoing evaluation.

Neurogenic bladder is common with tethered cord and after surgical repair of spinal dysraphism. Once neurogenic bladder occurs, reversal is unlikely. It is important to detect signs of early urinary dysfunction. Commonly a renal ultrasound and voiding cystourethrogram are performed as a baseline, followed by serial urodynamic evaluations.

Retethering of the spinal cord is common after surgical repair. Adhesions and scarring may occur where neural tissue touches the dorsal dura. Surgical techniques are being developed to assure circulation of CSF around the repaired cord, preventing contact with the dura.[32]

Terminal syringomyelia, a cystic dilation of the lower third of the spinal cord is associated with split-cord malformation and tethered cord. Dilation of the syringomyelia may increase after release of a tethered cord. It is thought that stretching of the cord may make the cyst thinner, and untethering allows the cord to shorten and dilate.[33]

Chiari II malformation and hydrocephalus is associated with nonterminal myelocystocele. Hindbrain abnormalities related to a smaller than normal posterior fossa are characteristic of this disorder.[24] See Part 2 of this series for a discussion of Chiari II malformation.[9]

Other problems include orthopedic issues, such as progressive muscle weakness and developmental abnormalities of the feet. Neurological dysfunction may lead to asymmetries in length of lower limbs, deformities of the feet such as reduced heel flexion or equinovarus, circumferential asymmetry, and asymmetry in foot size. Orthopedic disturbances are progressive.[4,18,34]

Scoliosis is common when a split-cord malformation is present or may occur if there is underlying vertebral abnormalities. Progressive scoliosis in the absence of vertebral anomalies usually reflects cord tethering.[10] A discussion of scoliosis will be the focus of Part 4 of this series on the spine.

Clinical Implications

The nurse's role in caring for infants with closed spinal dysraphism is to identify, describe, and communicate findings when cutaneous markings over the spine are present on physical examination. Monitor muscle tone and symmetry of the lower extremities. Weakness may be insidious and progressive or may be acutely precipitated by stretching the spinal cord (ie, with jackknife positioning).[10] Avoid extreme positioning for procedures such as lumbar puncture.

In the presence of cutaneous markings, lumbar puncture should not be performed until diagnostic imaging shows normal placement of the conus medullaris to avoid puncture of the spinal cord. Note that low-lying conus is not the same as a tethered cord but increases the risk of puncture in the same way.

Monitor and teach parents to watch for symptoms of urologic dysfunction because this is common with CSD. Symptoms may include urinary dribbling, overflow incontinence ("always wet"),[19] and frequent urinary tract infections. Symptoms often become apparent in retrospect. Explain the importance of urodynamic testing.[10,16,18]

Provide accurate information to the parents. Parents need to understand the importance of ongoing neurological follow-up after discharge. Coordinate follow-up with a developmental physician and a neurosurgeon at the time of discharge.

Summary

Closed spinal dysraphism poses challenges in identification because of the subtle appearance of some cutaneous markers and insidious onset of neurological and physical symptoms. Delayed diagnosis and intervention exposes the patient to multiple risks, including infection, increased tumor size, and the risk of developing neurologic deficit.[23] The trend toward earlier diagnosis and referral has made it possible for neurosurgical intervention to occur in a more timely manner and has decreased the development of complications in later life.[23] The nurse who is aware of the significance of midline cutaneous markers is in a position to facilitate the evaluation of potential CSD. Careful nursing assessment and documentation of the physical examination are important in identifying these lesions.

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