

## Neural Tube Defects

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### Developmental Anatomy and Sonography

#### The Neural tube

Between 5 & 6 weeks gestation (3-4 embryonic age), the neural tube forms from the neural plate. The embryo is 1-2 mm long at this stage. The rostral end of the neural tube becomes the brain and the caudal end becomes the spinal cord. The neural tube is temporally opened at both ends and communicates with the amniotic cavity. By 6 weeks gestation, both ends are closed, the lumen of the neural tube becomes the ventricular system of the brain and the central canal of the spinal cord. Surrounding the neural tube is the mesenchyme which developed into the dura matter and the bony spinal canal. At 10 weeks, the spinal cord extends the entire length of the vertebral canal & the spinal nerves exit the foramina at their levels of origin. Then, the dura matter and the bony spinal canal grow more quickly than the spinal cord. At 26 weeks the distal tip of the cord lies at S<sub>1</sub>. At full term it lies at L<sub>3</sub>. In the adult, it lies at L<sub>1</sub><sup>1</sup>.

#### Ossification of the Fetal Spine

The essence of sonographic evaluation of the spine is the evaluation of the bony spinal canal that surrounds the cord. The best visualized elements of the spine with perinatal sonography are the ossified portions: one in the centrum & two in the left and right processes. The centrum becomes the anterior central portion of the vertebral body. The neural process becomes the postero-lateral portion of the vertebral body. For the purpose of detection of spina bifida, ossification of the neural process is more important than the ossification of the centrum. Because neural tube defects manifest as abnormalities in the neural arches. Before 10 weeks gestation, the vertebrae are unossified cartilage. The ossification begins at 10 weeks in the centra & neural processes. At 13 weeks, foci of ossification are present in the neural process from C<sub>1</sub> to L<sub>5</sub>. From 13-20 weeks, ossification extends gradually into the pedicle & the lamina. By 18 weeks, enough ossification is present in the

pedicles and the laminae of the lumbar spine to effectively assess for spina bifida<sup>2</sup>.

#### Classification

The term neural tube defect refers to a group of malformations including spina bifida, anencephaly & cephaloceles.

#### Spina Bifida

##### Definitions

A midline defect of the vertebrae results in exposure of the contents of the neural canal. In the most majority of cases, the defect is localized to the posterior arch of the vertebrae. In rare cases, the defect consists of splitting of the vertebral body.

##### Incidence

Spina bifida is the most common malformation of the CNS. The incidence varies according to many factors (geographical, ethnic & seasonal)<sup>1, 2, 3, 4, 5</sup>

The spinal defects are more frequent in Caucasians than the Orientals or blacks. These differences seem to be persistent even after migration, suggesting a genetic rather than an environmental defect.

**Table [1]. Incidence of neural tube defects in different geographical areas**

	Incidence per 1000 births	
	SPINA BIFIDA	ANENCEPHALY
South wales <sup>5</sup>	4.1	3.5
Birmingham <sup>6</sup>	2.8	2.0
Alexandria <sup>7</sup>	2.0	3.6
Japan <sup>12</sup>	3.0	6.0

#### Etiology

Neural tube defects are most commonly inherited with a multifactorial pattern. They could also occur as a part of mendelian syndrome or chromosomal anomalies, or result from teratogenic exposure<sup>6, 7, 8</sup>.

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**Table [2]: Recognized causes of neural tube defects:**

<b>MULTIFACTORIAL INHERITANCE</b>	<b>Anencephaly, meningocele, meningomyelocele &amp; encephalocele</b>
<b>SINGLE MUTANT GENES</b>	<b>Meckel syndrome (AR)</b> Occipital encephalocele, rare anencephaly <b>Robert syndrome (AR)</b> Anterior encephalocele <b>Jarco Levin Syndrome (AR)</b> Meningomyelocele <b>HARDE Syndrome (AR)</b> Encephalocele
<b>CHROMOSOMAL ABNORMALITIES</b>	<b>Trisomy 13 , 18 &amp; Triploidy</b>
<b>TERATOGENS</b>	<b>Valproic acid:</b> Spina Bifida <b>Amnioplerin:</b> anencephaly & encephalocele
<b>MATERNAL PREDISPOSING FACTORS</b>	<b>DM:</b> Anencephaly more frequent than Spina Bifida

*Associated Anomalies*

The 2 main categories associated with spina bifida are other CNS defects and foot deformities. In almost all cases of spina bifida aperta, atypical abnormalities in the posterior fossa are found<sup>9</sup>. The lesion is Arnold Chiari malformation type 2, characterized by herniation of the Cerebellar vermis through the foramen magnum. The 4<sup>th</sup> ventricle is displaced downwards inside the neural canal. The posterior fossa is shallow and the tentorium is displaced downward. Displacement and kinking of the medulla are also observed. Arnold Chiari malformation is almost invariably associated with obstructed hydrocephalus<sup>10</sup>. The genesis of hydrocephalus seems to be related to the low position of the exit foramen of the 4<sup>th</sup> ventricle, which drains the CSF inside the spinal canal. Re-entry of the fluid to the intracranial cavity is then blocked by the cerebellum that obstructs the foramen magnum<sup>11, 12</sup>. In many cases deformities of the aqueduct are found, and these are believed to be secondary to ventricular enlargement & brain stem compression. Another frequently encountered CNS abnormality is polymicrogyria.

Dislocation of the hip & foot deformities in the form of club foot or rocker bottom foot is frequently seen in association with spina bifida. The pathogenesis of the malformation is related to the unopposed action of the muscle groups because of a defect of the peripheral nerve corresponding to the involved myotomes<sup>9</sup>.

*Diagnosis*

The criteria of the diagnosis of spina bifida are based upon soft tissue and bony signs. The soft tissue signs are "Absence of the skin covering the back, Presence of bulging sac". The bony signs are derived from the vertebral abnormalities associated with spina bifida. A clear understanding of the normal anatomy of the spine in different scanning planes is absolutely essential to the diagnosis. There are 3 main planes used in the evaluation of the spine: Sagittal, Coronal & Transverse which will be discussed later.

*Accuracy of the Ultrasound in Prenatal Diagnosis of Spina Bifida*

The detection of spina bifida is one of the most difficult tasks required of a sonographer. These examinations are known in the United States as level II scans and should be only be performed by very experienced operators.

The ultrasound is 94% sensitive & 98% specific for diagnosis of spina bifida when used in a population at risk (AF-AFP 3 SD above the

*Pathology*

Spina bifida lesions are subdivided into ventral and dorsal defects. The ventral defects are extremely rare and are characterized by the splitting of the vertebral body, and occurrence of a cyst that is neuroenteric in origin. The lesion is generally seen in the lower cervical or upper thoracic vertebra. Dorsal defects are most common and subdivided into spina bifida occulta & spina bifida aperta. Spina bifida occulta (15%) is characterized by small defect & completely covered by skin & in many cases it is asymptomatic and is diagnosed only incidentally by radiographic examination of the spine. Also, there is an area of hypertrichiosis, pigmented and dimpled skin, or the presence of subcutaneous lipoma. The clinical importance of this lesion is its frequent association with infection of the neural contents.

Spina bifida aperta (85%) is the most frequent lesion. The neural canal may be exposed, or the defect may be covered by a thin meningeal membrane. More often, the lesion appears as a cystic tumor (spina bifida cystica). If the mass contains purely meninges, the lesion is referred to as a Meningocele, or more frequently, neural tissue as a meningomyelocele<sup>1,6</sup>.

mean). The experience of the operator and the quality of the equipments are important factors in accurate perinatal diagnosis of these defects. However, a finite number of cases will not be diagnosable with sonography. Small sacral defects are probably the major diagnostic problem. The reason for this difficulty is that the integration of the sacral area is difficult because of its normal curvature and its flat shape.

**Prognosis**

Spina bifida is a serious congenital anomaly. The stillbirth rate is widely quoted to be 25%<sup>9</sup>. The majority of untreated infants die within the first few months of life<sup>13</sup>. Survival of infants treated in the early neonatal period is only 40% at 7 years<sup>13</sup>. Twenty five % of these infants are totally paralyzed, 25% are almost totally paralyzed, 25% require intense rehabilitation and only 25% have no significant lower limb dysfunction. Seventeen% of infants at late follow up have normal continence<sup>13</sup>. At present, it is impossible to predict while in utero the outcome of these infants. Prognostic factors include the level and the extent of the lesion and kyphoscoliosis. The presence of severe hydrocephalus has always been considered a poor prognostic sign<sup>13</sup>. Early neonatal shunting has significantly improved the intellectual development of these infants<sup>13, 14</sup>.

All infants of spina bifida have some degree of Arnold Chiari type II malformation. This condition is symptomatic (dyspnea, swallowing difficulties, opisthotonos), and represents a potentially fatal complication, only in a small number of cases. Death is usually related to respiratory failure. In a series, 45 infants with symptomatic Arnold Chiari malformation underwent laminectomy for relief of brain stem compression. The mortality rate was 38% in a follow up period ranging from 6 months to 6 years<sup>15</sup>.

**Obstetric Management**

The most important issue in obstetric management is the timing and mode of delivery. In the second trimester, the option of pregnancy termination should be offered to the parents. In the third trimester, parents should be counseled. Fetuses with spina bifida ideally should be delivered at term except those with rapid development of severe ventriculomegaly and macrocrania should be delivered earlier. There are inadequate data regarding the optimum route of delivery. The vaginal route could traumatize the defect and expose the neural tissue to bacteria normally present in the birth canal<sup>16, 17, 18, 19</sup>. For

this consideration it is better to deliver by cesarean section.

**Table [3]: Estimated incidence of neural tube defects based on specific risk factors in the United States:**

POPULATION	INCIDENCE PER 1000 LIVEBIRTHS
<b>MOTHER AS REFERENCE</b>	
General incidence	1.4 – 1.6
Women undergoing amniocentesis for advanced maternal age	1.5 – 3.0
Women with DM	20
Women with valproic acid in the 1 <sup>st</sup> trimester	10 – 20
<b>FETUS AS REFERENCE</b>	
1 <sup>st</sup> sibling with NTD	15 – 30
2 <sup>nd</sup> sibling with NTD	57
Parents with NTD	11
1/2 sibling with NTD	8
1 <sup>st</sup> cousin	10
Sibling with severe scoliosis secondary to multiple vertebral defects	15 - 30
Sibling with occult spina dysraphism	15-30

**Anencephaly**

**Definition**

Anencephaly is an anomaly characterized by the absence of cerebral hemispheres and cranial vault.

**Incidence**

The epidemiology of anencephaly is very similar to that of spina bifida [table1].

In neonates the anomaly is more frequent in females than males. The incidence of anencephaly in (abortions material) has been found to be five times greater than that observed at birth<sup>20</sup>.

**Etiology**

Anencephaly, as well as spina bifida has multifactorial etiology. A number of teratogenic agents including radiation, trypan blue, salicylates, sulfonamides<sup>21</sup>, and CO2 excess and anoxia<sup>22</sup> have induced this anomaly in experimental animals.

### **Pathology**

Most of the cranial vault is absent. The frontal bone is defective above the supraorbital region, and the parietal bones, as well as the squamous portion of the occipital bone are absent. The crown of the head is covered by the vascular membrane known as: "Area Cerebrovasculosa". Beneath the mass, few remnants of cerebral hemisphere can be found. The diencephalic and mesencephalic structures are either completely or partially destroyed. The hypophysis and the rhombo-encephalic structures are generally preserved. Other features that are characteristic to anencephalic infants include: bulging eyes, a large tongue and a very short neck.

### **Associated Malformations**

Spina bifida is present in 17% of patients (craniorachischisis), cleft lip or palate in 2%, club foot in 1.7%, omphalocele has also been described in some cases.

### **Diagnosis**

Anencephaly was the first congenital anomaly identified in utero with ultrasound. The diagnosis relies on the failure to demonstrate the cranial vault. Also has frog like appearance and usually have a short neck.

The diagnosis can probably be made as early as the 12-13 week.

In third trimester, the diagnosis is quite obvious when the fetus has transverse or breech presentation or while difficult when fetus is in vertex presentation. Because the base of the skull is often seen deep in the maternal pelvis.

Polyhydramnios is frequently associated with, anencephaly, the mechanism is unclear and several hypotheses have been suggested, including failure to swallow because of brain stem lesion, excessive micturition and failure of reabsorption of CSF.

Frequent accompanying phenomenon is increased fetal activity, may be due to meningeal irritation.

### **Prognosis**

The disease is uniformly fatal within the first hours or days. 53% are premature births, 15% postterm.

Only 32 percent of these fetuses are live births.

### **Obstetric Management**

Termination of pregnancy can be offered to the mother at anytime in gestation.

## **Cephalocele**

### **Synonyms**

Encephalocele, cranial or occipital meningocele and cranium bifidum.

### **Definition**

Cephalocele is a protrusion of the intracranial contents through a bony defect of the skull. The term "cranial meningocele" is used when only meninges are herniated. The term "encephalocele" defines the presence of brain tissue in the herniated sac. Encephalocele is commonly but incorrectly used to refer to both conditions.

### **Incidence**

Rare Occipital cephaloceles are by far the most frequent form in the Western World. In England, the frequency of this condition has been estimated to be 0.3 to 0.6 in 1000 births.

### **Etiology**

Other neural tube defects are often found in siblings in infants with cephalocele, implying a familial tendency. Besides the conditions associated with neural tube defects, cephaloceles are frequent components of a number of genetic syndromes (e.g., Meckel syndrome). They have also been reported in association with maternal rubella, diabetes and hyperthermia and can be produced experimentally in animals by the administration of several teratogens, such as X-ray radiation, trypan blue, and hypervitaminosis A<sup>23</sup>.

### **Embryology**

The basic disorder responsible for the defect is unknown. It has been suggested that overgrowth of the rostral portion of the neural tube may interfere with the closure of the skull. Alternatively, the defect may result from failure of closure induction by the mesoderm. Most cephaloceles are, therefore, located in the midline. An exception to this occurs in cases of amniotic band syndrome, in which cephaloceles may be multiple, irregular, or asymmetrical<sup>24, 25</sup>.

### **Pathology**

According to the bone in which the defect is located, cephaloceles are commonly subdivided into occipital, parietal, and frontal. By far the most common location is the occipital bone. The lesion may vary in size from a few millimeters to a mass larger than the cranial vault. It may contain only meninges (meningo-occele) or variable amounts of brain tissue (encephalocele).

In some cases, most of the brain tissue is contained in the herniated sac. Frontal cephaloceles occur more frequently between the frontal and ethmoidal bones (frontonasal cephalocele). Not all cephaloceles are externally evident<sup>26, 27</sup>. Some occur through a defect located in the base of the skull and protrude inside the orbits, nasopharynx, or oropharynx. Frontal cephaloceles almost always contain brain tissue.

**Association anomalies**

As previously mentioned, cephaloceles can be found as part of a number of specific syndromes. In addition, both meningoceles and encephaloceles are associated with other CNS abnormalities. Hydrocephalus has been reported in 80 percent of occipital meningoceles, 65 percent of occipital encephaloceles, and 15 percent of frontal cephaloceles. Spina bifida is found in 7<sup>28, 29, 30</sup> to 15 percent of all cephaloceles<sup>10</sup>. Microcephaly was observed in 20 percent of cases studied. By definition, the herniation of the cerebellum inside the cephalocele is termed "Chiari type III deformity". This deformity, combined with aqueductal stenosis, is the major cause of hydrocephalus in these infants. Frontal cephaloceles are often associated with the median cleft face syndrome, characterized by hypertelorism and median cleft lip or palate<sup>31</sup>.

**Diagnosis**

Traditionally, the diagnosis of cephalocele relies on the demonstration of a paracranial mass. However, this criterion is insufficient to distinguish them from other non neural masses, such as cystic hygromas, and soft tissue masses, such as scalp edema. For this reason, an effort should be made to identify the skull defect is usually smaller than the herniated mass and sometimes falls below the resolute power of current ultrasound equipment. In axial scan, the complete controller of the occipital and frontal bones is not adequately visualized because of sound refraction. Furthermore, the normal sutures can be confused with a defect<sup>32, 33, 34</sup>.

**Hints for a proper differential diagnosis are:**

- Cephaloceles are often associated with hydrocephaly.
- Brain tissue can be seen in some cephaloceles.
- Cystic hygromas usually have multiple septa, are often associated with other signs of hydrops, and have a paracervical origin.

- Severe scalp edema can be confused with a cephalocele, but usually a sagittal scan can identify an intact skull and the diffuse nature of the condition.

**Table [4]. Conditions associated with cephaloceles**

<p><b>Amniotic band syndrome (sporadic)</b> Multiple cephaloels, predominantly anterior Amputations of digits or limbs Bizarre oral clefts</p> <p><b>Cryptophthalmos syndrome (AR)</b> Forehead skin covers one or both eyes Ear abnormalities Soft tissue syndactyly</p> <p><b>Dyssegmental dysplasia (AR)</b> Short limb dysplasia Metaphyseal widening Small thorax Micrognathia</p> <p><b>Frontonasal dysplasia (sporadic, some cases are familial)</b> Frontal cephalocele Ocular hypertelorism</p> <p><b>Meckel syndrome (AR)</b> Polycystic kidneys Polydactyly Microphthalmia Orofacial clefting Ambiguous genitalia</p> <p><b>Von Voss syndrome</b> Agenesis of the corpus callosum Phocomelia Urogenital anomalies Thrombocytopenia</p> <p><b>Warfarin syndrome</b> Nasal hypoplasia Bone stippling Limb shortening Hydrocephaly</p> <p><b>Associations</b> Absence of corpus callosum Cleft lip or palate Craniostenosis Dandy-Walker syndrome Ectrodactyly Hemifacial microsomia Iniencephaly Meningomyelocele</p>
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Amniotic fluid alpha-fetoprotein (AFAFP) is usually elevated when the brain tissue or meninges are exposed. However, we have seen one case with a defect covered by skin in which the level of AFAFP was normal.

Whenever the diagnosis of a cephalocele is made, a careful examination of the fetus is indicated to search for other associated anomalies<sup>35</sup>.

### **Prognosis**

The prognosis of cephaloceles depends on three factors

- The presence of brain in the herniated sac.
- Hydrocephalus.
- Microcephaly.

The most important prognostic factor is the herniation of the brain. The mortality rate in these cases has been reported to be 44 percent versus no deaths observed in cases of simple meningocele.<sup>10</sup> Intellectual development was normal in only 9 percent of patients in the former group and 60 percent in the latter.<sup>36, 37, 38</sup>

The "influence" of hydrocephalus on intellectual development is controversial. Lorber observed no significant difference in the groups of infants with arid without ventriculomegaly. In another series reported by Guthkelch, 86 percent of patients with meningocele without hydrocephalus had an IQ higher than 70, whereas only 50 percent of those with hydrocephalus had IQs above this level.

The effect of microcephaly has been reported in a limited number of infants. Lorber observed that 3 of 8 infants with microcephaly died, and the remaining five were intellectually impaired.

### **Obstetric Management**

Termination of pregnancy should be offered before viability. In the third trimester, obstetrical management depends on the size of the defect, the amount of herniated brain tissue, and associated anomalies. If associated anomalies incompatible with life are present (e.g. Meckel syndrome), termination in the third trimester can be undertaken. In the absence of such findings, patients should be counseled. Theoretically, a cesarean section could improve prognosis by avoiding birth trauma and contamination of brain tissue with vaginal flora. Non-aggressive management is recommended in case of microcephaly.

### **Scanning Techniques**

#### **Optimal scan planes**

The most useful scan planes to evaluate the posterior spinal arches are

- Posterior transaxial.
- Lateral transaxial.
- Lateral longitudinal.
- Posterior longitudinal.

The posterior transaxial scan plane is optimal for the laminae and posterior skin surface. The lateral transaxial plane is optimal for the pedicles. The posterior longitudinal plane is excellent for the posterior skin surface detection of a myelomeningocele sac. The lateral longitudinal scan plane allows direct comparison of interpedicular distances at multiple levels<sup>39, 40</sup>.

#### **Routine complete sonogram (level 1 sonogram)**

Sonograms to assess curly fetal anatomy are best performed at 18 weeks gestation. The scanning techniques described here are best suited for this age, but may also be used at other stages of pregnancy. To start, the sonographer must determine the position of the fetal head and spine. Then the fetal head is scanned in standardized<sup>39, 40, 41</sup> scan planes because most fetuses with spina bifida have head abnormalities at 18 weeks gestation: obliterated cisterna magna (banana sign) and/or enlarged lateral ventricles and/or concave frontal bones (lemon sign). If one or more of the head abnormalities is present, a detailed scan of the spine is indicated.

In fetuses with open neural tube defect between 16 and 24 weeks gestation, the posterior fossa abnormalities are present in 95% to 100% (3-5), ventriculomegaly in 44% to 86%. (3-6) and the lemon sign in 85% to 100% (4.7.8). In contrast, experienced sonographers detect only 80% to 90% of the actual spinal abnormalities in fetuses with spina bifida (5).

For a routine sonogram of the fetal spine (i.e., no risk factors in the pregnancy, no fetal head abnormality identified), the following are suggested:

- Place the scan plane perpendicular to the long axis of the fetal spine (posterior transaxial or lateral transaxial).
- Scan the entire spine while maintaining the scan plane perpendicular to the spinal long axis no matter how the spine is curved.
- Scan through the entire spine several times quickly in this fashion: this builds up a three-dimensional image of the spine from the cervical spine to the sacral spine.
- Reposition the scan plane parallel to the long axis of the spine to obtain posterior

longitudinal or lateral longitudinal scans of the spine from the cervical spine to the sacral spine<sup>42, 43, 44, 45</sup>.

### ***Detailed Sonogram (Level II or Targeted Sonogram)***

A more detailed sonogram of the spine may be required for one of several reasons:

- Head abnormality detected in current sonogram.
- Possible spinal abnormality detected in previous sonogram.
- Recognized risk factors.
- Raised maternal serum AFP (positive Maternal Serum Screen Test).

A detailed sonogram often requires more time by the sonologist to evaluate each level of the spine with as many scan planes, probes, machines, and maneuvers that are required for absolute optimization. This could include endovaginal scans or repeat scans at a later date if fetal position is suboptimal in the current sonogram<sup>44, 45, 46, 47</sup>.

### **Artifacts, Pitfalls, and Practical TIPS**

#### ***Open Versus Closed Neural Tube Defects***

An open defect implies there is no skin cover. Most myelomeningocele sacs are covered by only a meningeal membrane without skin and are thus classified as "open." Open defects have elevated AFP levels whereas closed (i.e., skin-covered) defects have normal AFP levels.

#### ***Unossified Coccyx Simulating Abnormality***

The lower sacrum and coccyx in mid-trimester are echo-free cartilaginous structures. A posterior longitudinal sonogram of the coccyx simulates a fluid-filled tract. This can cause confusion in detailed sonograms of the lower spine and may simulate an abnormal communication between a neighboring mass (e.g., sacrococcygeal teratoma) and the spine<sup>47, 48</sup>.

### **False Negative Spina Bifida**

#### ***Oligohydramnios***

With oligohydramnios, all fetal structures including the fetal spine are more difficult to visualize and thus spina bifida defects may be missed. With oligohydramnios, the wall of a myelomeningocele sac is pushed against the wall of the uterus, and thus the sac is much more difficult to appreciate. In this circumstance, one must very carefully examine the ossified laminae and pedicles because the myelomeningocele sac may be obscured. Severe oligohydramnios may

also cause positional curvatures in the fetal spine and thus divert attention away from a site of spina bifida<sup>49</sup>.

#### ***Spina Bifida without a Sac***

Spina bifida is easier to detect if a myelomeningocele sac or meningocele sac is present. Therefore, one must seek not only a posterior sac, but, more importantly, an abnormality of the ossified laminae and pedicles of the fetal spine<sup>50, 51</sup>.

#### ***Small Defect in Midline***

A narrow spina bifida defect in the midline posteriorly may not be associated with a detectable abnormality of the ossified portions of the laminae. Posterior longitudinal and posterior transaxial scans may detect a small defect in the nonossified soft tissues, but this can be easily overlooked unless there is an accompanying cystic mass.

#### ***Scan Too Early in Pregnancy***<sup>50, 51, 52, 53</sup>

Before 16 to 18 weeks gestation, relatively extensive spina bifida may be missed for two main reasons:

- The structures are sufficiently small that the ultrasound resolution is incapable of depicting the abnormalities.
- Spinal ossification has not progressed enough to allow appreciation of the spina bifida defect.

Therefore, reports from early fetal sonograms should indicate that spinal evaluation is suboptimal compared to later in pregnancy. Evaluation is best performed at 18 weeks gestation or later. Note, however, that spina bifida can certainly be detected in early pregnancy (even first trimester with optimal scanning conditions: high resolution systems, endovaginal probes, fetal spine close to the ultrasound probe<sup>54, 55</sup>).

#### ***Suboptimal Scan Planes***

The optimal scan planes for spinal assessment are the posterior longitudinal and posterior transaxial planes: the best fetal position is prone. If the fetus is supine, adequate assessment is usually not possible. If the fetus is lateral decubitus, adequate assessment is usually possible: lateral longitudinal and lateral transaxial scan planes are used for spinal assessment.

If oblique longitudinal or angled transaxial scan planes are obtained, erroneous impressions may result. If the longitudinal scan plane intersects the laminae on one side of the spine and the vertebral bodies on the other side these ossification centers

may be mistaken for the left and right pedicles. If the lateral longitudinal scan plane is too anteriorly located, the ossified centers and the left and right pedicles create a series of three parallel structures, which may cause confusion<sup>54, 55</sup>.

### False Positive Spina Bifida

#### *Normal Widening of the Interpedicular Distance*

In normal fetuses, the interpedicular distances in the cervical spine are significantly larger than the thoracic spine. The lumbar interpedicular distances are also slightly greater than the lower thoracic spine (by 1 to 2mm). These normal findings may cause concern to the less experienced examiner. All other findings in the spine will be normal.

#### *False Positive Maternal Screen for Spina Bifida (Wrong Dates)*

A common cause for a "positive" serum screen test is wrong dates, in the LMP gives an estimated gestational age significantly less than true dates, the laboratory will interpret the AFP level as abnormally high. The subsequent detailed fetal sonogram will establish the correct dates and hopefully correct the misconception. The error may be avoided if all women first have a dating sonogram before the maternal screen test, which is drawn ideally at 16 weeks gestation.

#### *Sacroccygeal Teratoma (SCT)*

An SCT with external component in the buttock area may simulate a myelomeningocele sac, but the location is usually more caudal than lumbar or sacral spina bifida. In addition, the defects in the laminae and pedicles are usually detectable in myelomeningocele, whereas the laminae and pedicles are normal in SCT. However, the unossified coccyx may simulate a communication between the SCT and the spine. An awareness of the normal appearance of the coccyx will avoid confusion<sup>56-59</sup>.

#### *Nuchal Masses*

Cystic hygromas are the most common posterior nuchal mass and they may simulate a myelomeningocele, especially if the examiner is less experienced. If the ossified spinal elements are normal and (the brain is normal, the likelihood of an NTD is very low. The one exception is a skin-covered cervical meningocele or myelocystocle, where the brain and ossified elements may be normal on ultrasound. However, these masses are often more or less spherical and

midline, whereas cystic hygroma usually extend more laterally.

#### *Encephalocele*

A posterior encephalocele may be impossible to distinguish from a high occipital myelomeningocele, although the distinction may not be critical for clinical management. An encephalocele and cervical myelomeningocele may coexist and be continuous with one another. With iniencephaly, the cervical spine is shorter than normal and there is abnormal extension of the fetal head and cervical spine<sup>60-62</sup>.

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