MICHIGAN MEDICINE
UNIVERSITY OF MICHIGAN

Fetal Myelomeningocele (MMC) (Spina Bifida) Glossary

Fetal Myelomeningocele (Fetal MMC) is a defect in the spine and spinal cord that occurs when a baby is still in the womb (a fetus). Fetal MMC occurs when the spine, spinal cord and spinal canal don't close into a "tube" (spinal cord) as they normally would. It is a serious form of **spina bifida** (where the spine never completely wraps around the spinal cord, leaving an opening in the spine).

The terms below are included as a glossary that will help you understand the Stickman illustration on the last page. These are the common birth defects seen in infants with Fetal MMC.

Chiari II hindbrain herniation: A condition that affects how the back of the brain is formed. The back of the brain is positioned farther down into the upper spinal column because of the Fetal MMC defect. This blocks the normal flow of fluid out of the brain and may be associated with:

- Hydrocephalus-occurs when there's too much cerebrospinal fluid in the head.

Or in more severe cases:

- Breathing to stop (Apnea) (blue spells)
- Low heart rate (bradycardia)
- Choking (Aspiration) and repeated lung infection (pneumonia)

Corpus callosum deficiencies: These are birth defects in which there are problems with the connection between the left and the right side of the brain (termed the corpus callosum). Abnormalities in the corpus callosum can be associated with developmental delays (see definition below).

CT Scan: Computed tomography, called CT or CAT scanning, is an x-ray system that uses a computer to take detailed images of areas of the body. These pictures look like cross-sections or "slices" of the body that are then put together by the computer. Some CT scans require the use of a contrast material,

sometimes called “contrast dye” or “iodinated contrast material”, which involves radiation exposure.

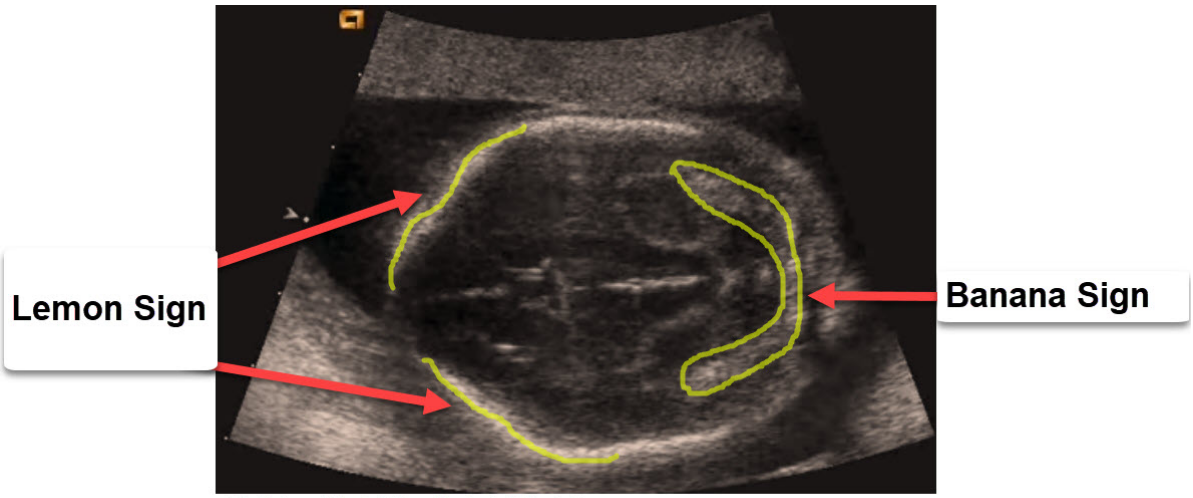
Developmental delay:
Developmental delay refers to a child who has not gained the developmental skills expected of him or her, compared to others of the same age. Delays may occur in the areas of motor, speech and language, cognitive, play and social skills.

Incontinence: Problems with the S-2, S-3, S-4 nerves can cause issue with the bowel and bladder holding urine.

Lemon and banana signs refer to the shape of the head on an ultrasound and can be indicators of Fetal Myelomeningocele.

- **Lemon Sign:** is when the bones in the front of the skull are sunken in, or flattened instead of rounded. This gives the front of the scull a lemon shape. A lemon sign doesn’t always mean spina bifida. Some babies don’t show other symptoms of Fetal MMC but may still have the lemon sign.
- **Banana sign:** refers to the shape of the cerebellum, a major part of the back of the brain.

The image below shows what the lemon and banana sign might look like on a brain scan.



This Photo was taken by Wolfgang Moroder via Wikimedia Commons: https://commons.wikimedia.org/wiki/File:Lemon_and_banana_sign_for_neural_tube_defect_Dr_Wolfgang_Moroder.theora.ogv. Modified screenshot from video to include highlighted areas and add labels and arrows.

MRI Scan: A procedure in which radio waves and a powerful magnet linked to a computer are used to create detailed pictures of areas inside the body. These pictures can show the difference between normal and abnormal tissue. MRI makes better images of organs and soft tissue than other scanning techniques, such as computed tomography (CT) or x-ray. Also called magnetic resonance imaging, NMRI, and nuclear magnetic resonance imaging. MRI does not use radiation to make the images and is considered safe in pregnancy.

Scoliosis is when the spine is curved from side to side and shaped like the letter S or C.

Shunts (a drainage for the extra fluid around the brain**)**:

- Obstruction: a blockage of the drainage tube
- Displacement: the tube moves and is no longer working
- Infections

Sexual function

- Spina Bifida patients with lesions at, or below S1 level typically have minimal neurological (brain) defects and may have sexual function similar to people without spina bifida.
- Spina Bifida lesions between L3-L5 will have different sensations than those without spina bifida and men may need assistance to father children.

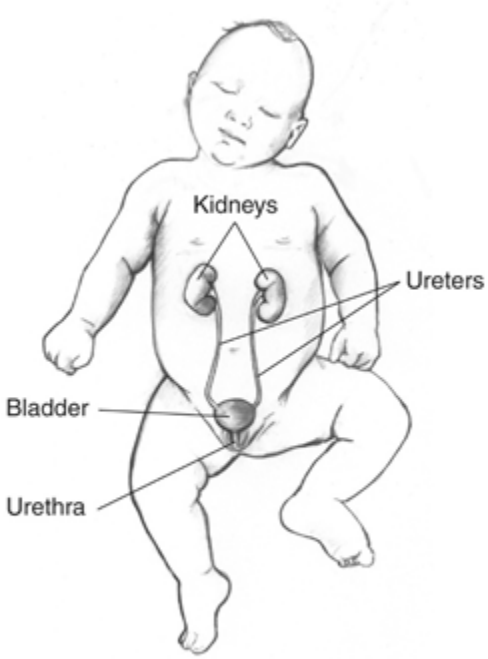
Talipes equinovarus (clubfoot deformity)- is a deformity of the foot present at birth that impacts one or both feet. A child with clubfoot has an abnormally positioned foot that causes the child to turn their ankle inward, to the point where it almost touches the inside of the leg.

Ultrasound Scan: An ultrasound is a procedure that uses high-energy sound waves to look at tissues and organs inside the body. The sound waves make echoes that form pictures of the tissues and organs on a computer screen (sonogram). **Echoes** are sound waves that are reflected or bounced off different tissues. Different tissues interact with sound in different ways so normal tissue, abnormal tissue and defects in tissue can all be identified with ultrasounds. Ultrasound is considered safe in pregnancy.

Urinary reflux occurs if the valve fails where the ureter meets the bladder. This causes urine from the bladder to back up into the ureter and kidney. This can cause:

- Repeated kidney infections.
- Kidney failure and the possibility of dialysis and a kidney transplant.

Urinary stasis, also called urinary retention, is when the bladder cannot empty completely. To urinate, signals from your brain have to travel through your spinal cord and surrounding nerves then to your bladder and sphincters. If one or more of these nerve signals doesn't work, it can cause urinary stasis.



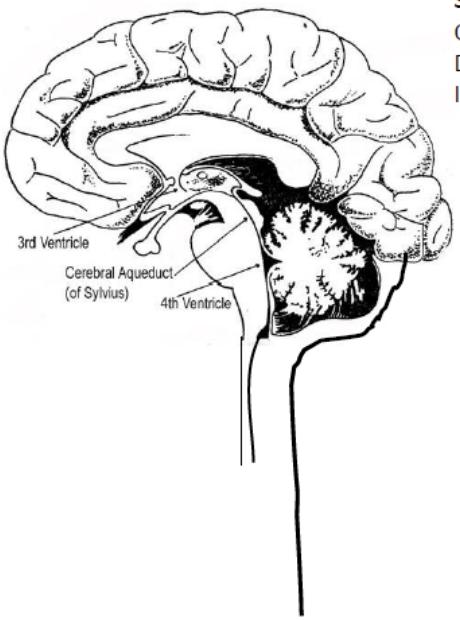
National Institute of Diabetes and Digestive and Kidney Diseases, National Institutes of Health.

Genetics

Multifactorial inheritance: Many factors (multifactorial) are involved in causing a Fetal MMC birth defect. The factors are usually both genetic and environmental. This means a combination of genes from both parents, in addition to unknown environmental factors, produce the condition.

Recurrence risk is the chance that an inherited disease that is present in a family will reappear in that family, affecting another person. The recurrence risk for Fetal MMC is 4-6%.

- Taking 4-5 grams of folic acid (folate) daily starting at least 3 months before your next pregnancy decreases risk to 1%.



Shunts:
Obstruction
Displacement
Infections

" Lemon Sign"

Corpus callosum deficiences

Hydrocephaly

"Banana Sign"
Chiari II hindbrain herniation
↓
Posterior fossa compression

*Apnea (blue spells)
*Bradycardia (low heart rate)
*Aspiration
Choking
Repeated pneumonias

Risk for scoliosis

Urinary stasis and reflux
↓
Recurrent kidney infections
↓
Renal failure, dialysis, transplant

S-2, S-3, S-4: bowel and bladder incontinence

L-1, L-2

sexual function

L-3, L-4

L-4, L-5, S-1, S-2

Talipes equinovarus
(clubfoot deformity)

Genetics
*Multifactorial inheritance
*Recurrence risk 4-6%

Folic acid supplement (folate)
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