Tuesday, February 12, 8:00AM-9:50AM
Plenary Session

Defects of the Ventral Wall and Related Complexes
Moderator: Angela Lin, Massachusetts General Hospital, Boston, MA

Embryology of Ventral Wall Defects
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During the 3rd and 4th weeks post-fertilization (5 and 6 weeks from the LNMP), the human embryo is transformed from a flat disc-shaped organism into the classic shape of an embryo in the fetal position. This change is effected by simultaneously rolling the top layer of the disc, the ectoderm, into the neural tube and the bottom layers of the disc, the endoderm and mesoderm, into the gut tube and body wall, respectively. The body wall is formed by ventrally moving lateral body wall folds that are a combination of mesoderm and ectoderm that arise on each side of the embryo. Forces that move the folds ventrally include proliferation of cells in the existing mesoderm and ectoderm layers and migration of new mesoderm from adjacent tissues (somites) into the folds. By the middle of the fourth week, the folds approach each other in the midline and make contact. At this time, cell death and migration must occur such that appropriate tissues (amnion, mesoderm, and ectoderm) fuse and establish continuity from side to side. Also, at this time, surface coats, which act like glue, must be synthesized on the leading edges of each fold immediately prior to contact. Assisting this process are the head and tail folds that cause the embryo to curve into the fetal position. However, these folds are formed in a more passive manner and probably do not play a major role in actual closure of the ventral body wall. Closure itself is completed by the end of the fourth week.

Ventral body wall defects probably occur in much the same way that neural tube defects (NTDs) arise. For example, it is known that if the neural folds, which resemble lateral body wall folds, fail to elevate, then an NTD results. Failure of elevation may involve cell proliferation in the ectoderm or mesoderm or abnormal deposition of extracellular matrices. Perhaps similar events are involved in ventral body wall defects. Furthermore, surface coats (glue) are essential for neural fold closure and their disruption leads to NTDs. Again this same phenomenon may be involved in body wall closure. In any case, if body wall closure fails in the thorax, ectopia cordis results; if it fails in the abdomen, then it results in gastroschisis or Cantrell pentalogy if it is a large abnormality; and if it fails in the pelvis, then the defects that occur are bladder and cloacal extrophy, depending on the severity of the abnormality. Interestingly, no known closure defects have been described for the gut tube despite the fact that it too involves folding of a flat layer of cells into a tube. Note, however, that all of the closure defects that do occur have their origins early in gestation during the third to fourth weeks of development.