

Tuesday, February 12, 10:10AM-11:45AM
Plenary Session

Defects of the Ventral Wall and Related Complexes

Moderator: Angela Lin, Massachusetts General Hospital, Boston, MA

Amniotic Band Syndrome and Limb-Body Wall Complex

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The amniotic band syndrome (ABS) is a phenotypically diverse pattern of congenital anomalies that, in some cases, results from early rupture of the amnion; this leads to the formation of mesodermal fibrous strands that adhere to and/or entangle fetal structures, causing deformations and disruptions. Alternatively, ABS has been attributed to severe hypoxia and/or a genetic predisposition leading to vascular disruption and resulting in secondary attachment of fibrous bands to abnormal tissue. There is no clear consensus as to the pathogenesis of ABS, and multiple mechanisms may, in fact, be involved. Various patterns of generally asymmetric anomalies are attributed to ABS: (1) isolated limb or digital band constrictions, syndactylies or amputations; (2) craniofacial disruptions +/- limb anomalies; and (3) body-wall complex (BWC) +/- limb anomalies and/or craniofacial disruptions. The incidence of ABS has been reported in more recent studies to vary between 1:1,230 and 1:10,160 births. The incidences in preivable fetuses and spontaneous miscarriages have been reported as even more common, at approximately 1:56 and 1:250, respectively. The majority of cases occur sporadically, although rare familial cases have been reported. Many maternal and prenatal factors have been reported in previous epidemiologic studies or case series to be associated with ABS, although only young maternal age and primiparity have been associated with ABS in more than one epidemiologic study. These studies have been limited by small numbers of cases. The goals of this presentation will be to (1) review the potential mechanisms for ABS and limb-body wall complex (LBWC) in relation to fetal embryology, (2) describe the results of a population-based survey of the clinical phenotype of ABS and LBWC, and (3) examine the relationship between potential risk factors and the occurrence of different ABS patterns.