Microgastria-Limb Reduction Defects Association

Alternative Names
MLRD

WHO International Classification of Diseases
Congenital malformations, deformations and chromosomal abnormalities

OMIM Number
156810

Mode of Inheritance
Isolated cases

Description
Microgastria-limb reduction defect is a rare disorder of unknown etiology. Most of the cases of congenital microgastria reported in the literature are associated with other multiple congenital anomalies like; limb-reduction defects, asplenia, intestinal malrotation, hepatic symmetry, cardiopulmonary anomalies, central nervous system and renal anomalies, and laryngo-tracheo-bronchial clefts. Isolated congenital microgastria is an extremely rare condition. The microgastria-limb reduction association is believed to arise as a result of abnormal mesodermal development in the fourth or fifth week of embryonic life. All cases of microgastria associated with limb reduction defect have been sporadic with equal male: female distribution.

In the absence of other associated life-threatening congenital anomalies, microgastria can be managed successfully with early gastric augmentation leading to toleration of increasing amounts of oral feeding.

Molecular Genetics
There is no evidence of Mendelian inheritance of microgastria-limb reduction defect. The occurrence of twinning in three out of 13 cases suggested that the origin of the microgastria-limb reduction complex may be related to the process of twinning itself.

Epidemiology in the Arab World

Sudan
Al-Gazali et al. (1999) studied two Sudanese patients, residents of the UAE, with microgastria-limb reduction defect with central nervous system anomalies. The two cases were brothers resulting from a first-cousin marriage. There were five other normal children, and the family had no history of any abnormalities. Both cases showed multiple congenital abnormalities, which were detected by using skeletal survey, CT scan, and ultrasonography. The abnormalities included: bilateral microphthalmia, dysmorphic ears with an atretic auditory canal in one of them, bilateral radial aplasia with absent thumbs, oligodactyly, esophageal and anal atresia, abnormal kidneys and dilated lateral ventricles with agenesis of corpus callosum. Chromosome analysis was normal for the two cases. The first infant had, in addition, hemivertebra and sacral agenesis. He was a product of a complicated pregnancy with polyhydramnios. He died at 19 days of age. On the other hand, the pregnancy of the second neonate was complicated by gestational diabetes, and he died at 2 days of age. The recurrence of this condition in siblings of these consanguineous parents supported autosomal recessive inheritance in this subgroup. Those two cases of Al-Gazali et al. (1999) raised the number of cases in which there is microgastria, limb reduction defect, CNS anomalies, and other congenital abnormalities to six reported cases worldwide.

United Arab Emirates
See: Sudan > Al-Gazali et al., 1999.

References
Al-Gazali LI, Bakir M, Dawodu A, Nath R, al-Tatari HM, Gerami M. Recurrence of the severe form of microgastria-limb reduction

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