



Adams-Oliver Syndrome

Alternative Names

Absence Defect of Limbs, Scalp, and Skull
AOS

Aplasia Cutis Congenita with Terminal Transverse
Limb Defects

Aplasia Cutis Congenita, Congenital Heart Defect,
and Frontonasal Cysts

Congenital Scalp Defects with Distal Limb Reduction
Anomalies

Record Category

Disease phenotype

WHO-ICD

Congenital malformations, deformations and
chromosomal abnormalities > Other congenital
malformations

Incidence per 100,000 Live Births

Unknown

OMIM Number

100300

Mode of Inheritance

Autosomal dominant, and autosomal recessive

Gene Map Locus

N/A

Description

Adams-Oliver syndrome is a rare congenital anomaly complex characterized by vertex scalp defect like aplasia cutis congenita, terminal transverse defect of limbs and cutis marmorata of skin. Hand deformities include postaxial polydactyly, short fingers or absent hand, phalanges, or digits and syndactyly. Foot deformities include short toes, absent foot, distal phalanges, toes or feet, widely spaced toes, clubfoot, and dermatoglyphic abnormalities.

Molecular Genetics

Adams-Oliver syndrome is inherited as autosomal dominant with marked variability in expression.

Epidemiology in the Arab World

United Arab Emirates

Al Talabani et al. (1998) studied the pattern of major congenital malformations in 24,233 consecutive live and stillbirth in Corniche hospital, which is the only maternity hospital in Abu Dhabi, between January 1992 to January 1995. A total of 401 babies (16.6/1,000), including 289 Arabs, were seen with major malformation. Single gene disorders accounted for 24% of the cases, 21% were due to autosomal dominant disorders. In their study, Al Talabani et al. (1998) observed one case of Adams-Oliver syndrome in a family from the United Arab Emirates. Recurrence was reported in the family. Al Talabani et al. (1998) concluded that this study is very close to representing the true incidence of congenital abnormalities in the whole United Arab Emirates, as this study included over 98% of deliveries in Abu Dhabi, the capital of United Arab Emirates.

References

Al Talabani J, Shubbar AI, Mustafa KE. Major congenital malformations in United Arab Emirates (UAE): need for genetic counselling. *Ann Hum Genet.* 1998; 62 (Pt 5):411-8. PMID: 10088038

Related CTGA Records

N/A

External Links

<http://www.cafamily.org.uk/Direct/a18.html>

<http://www.emedicine.com/derm/topic32.htm>

http://www.rarediseases.org/search/rdbdetail_abstract.html?disname=Adams%20Oliver%20Syndrome

Contributors

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