IS-019
INCREASED LEVELS OF CIRCULATING ADHESION MOLECULES IN NEONATES WITH
CONGENITAL DIAPHRAGMATIC HERNIA COMPLICATED BY PERSISTENT PULMONARY
HYPERTENSION

Department of Pediatric Surgery, Juntendo University School of Medicine a), Childrens Research Centre, Our Ladys Hosp-
ital for Sick Children b)
Hiroyuki Kobayashi b), Atsuyuki Yamataka b), Puri Prem b), Miyano Takeshi b)

Purpose: To investigate the relationship between soluble ICAM-1, ELAM-1 and VCAM-1 from patients with congenital
diaphragmatic hernia (CDH) and the development of persistent pulmonary hypertension (PPH).

Methods: We measured serum levels of adhesion molecules in twenty neonates with CDH at the time of diagnosis:9 with
PPH and 11 without PPH) and 7 controls using ELISA. We further examined the lungs with CDH complicated by PPH who
died. and three control for the expression of adhesion molecules using immunohistochemistry.

Results: The mean serum ELAM-1 (116.5±19.2Mg/ml), VCAM-1 (1596.9±60.4Mg/ml) and ICAM-1 (227.0±98.9Mg/ml) levels
in CDH patients with PPH were significantly increased compared to the levels in CDH patients without PPH(ELAM-
1 (79.3±27.9Mg/ml), VCAM-1 (1069.3±444.6Mg/ml) and ICAM-1 (140.2±93.7Mg/ml)) and controls. But serum adhesion
molecules levels in CDH patients without PPH were no different from controls statistically.

Pulmonary vascular endothelial cells of the CDH lung with PPH expressed strong adhesion molecules compared to con-
trols.

Conclusions: Upregulated expression of adhesion molecules on the endothelial of pulmonary vasculature and high cir-
culating levels of adhesion molecules in CDH patients with PPH suggest that adhesion molecule may play a role in the
development of PPH.

IS-020
Recent Advances in Congenital Diaphragmatic Hernia

Department of Paediatric Surgery, Institute of Child Health
Royal Liverpool Children's Hospital ( Alder Hey ) & The University of Liverpool, UK
Paul D. Losty

Congenital diaphragmatic hernia (CDH) is a lethal birth defect with an incidence of 1:2,500 births. A baby is born every 24-
36 hrs in the UK with CDH. Pulmonary hypoplasia and pulmonary hypertension account for the persistently high mortality
that claims the lives of almost 35%-45% of newborns. In utero deaths are responsible for a substantial “hidden mortality”.努力
Efforts to improve survival have focused on fetal surgery, in utero diagnosis with delivery at specialist centres, advances in
intensive care medicine ( iNO, ECMO ) and delayed surgical repair. Developmental biology studies have led physicians and
surgeons to appreciate that CDH is now not just “a hole in the diaphragm”. Traditionalist compression theory on the origins
of lung hypoplasia in CDH have been challenged by experimental studies of primordial lung development. Crucially such
studies have indicated a primary disturbance in lung organogenesis that precedes closure of the diaphragmatic defect.
Organotypic culture systems are providing invaluable research tools to investigate pulmonary hypoplasia and the role of
growth factor(s) / cell signalling pathways. Fetal surgery (FETENDO) based on the PLUG concept may be “too little too late”
to correct an established defect in embryonic lung development. Reliable indicators of prenatal lung growth to aid case
selection of the “high risk” CDH fetus are needed. Pharmacological strategies to ameliorate pulmonary hypoplasia are
under investigation. Antenatal corticosteroid therapy is the subject of an international multicentre trial. Postnatal therapies to
reduce ventilator-induced lung injury (permissive hypercapnia) and liquid ventilation hold future promise. Further advances
may arise from improved understanding of primitive events regulating airway physiology-lung growth. Preconceptual
prophylaxis ( as in the case for neural tube defects ) may be the final solution for this highly lethal human anomaly.