P-005  Congenital Diaphragmatic Hernia-Scoring Form
Al Wasl Hospital, Department of Health and Medical Services, Dubai, United Arab Emirates
Al Wasl Hospital, Department of Health and Medical Services, Dubai, United Arab Emirates
Hasan Nougoud, Abdul Rahim Mustafawi

A 7 year (Jan/2001 to Dec/2006) study was done to review and audit our cases of CDH. Purpose: The purpose of study was to establish a protocol for selection of neonates for proper intervention timing. Methods: Retrospective Study. 50 cases were thoroughly analyzed. A special form was developed for this purpose. Data were introduced by a professional multi-disciplinary team of neonatologist, paediatric anaesthetist, paediatric surgeon and SCBU well trained nurses. Summary of Results: The scores were compared to the thus taken decisions for every case and the outcomes were compared for auditing purposes. The results were discussed. Conclusion: The conclusions and outcome supported the form to be used prospectively for all cases of CDH to help classifying the patient for the proper mode of management.

P-006  Esophageal atresia associated with coarctation of the aorta, CHARGE association, and DiGeorge syndrome: A case report
Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, Tokyo, Japan
Kyeong Deok Lee, Tadaharu Okazaki, Yoshifumi Kato, Geoffrey J Lane, Atsuyuki Yamataka

Aim: We report an extremely rare case of esophageal atresia and tracheo-esophageal fistula (TEF) associated with coarctation of the aorta (CoA), CHARGE association, and DiGeorge syndrome.

Case report: A boy born by cesarean section at 38 weeks gestation, birth weight 2950 g, presented elsewhere with severe respiratory distress and was transferred to our institution. On admission, the boy had a peculiar appearance and Gross type C TEF and CoA. Gastrostomy was created on day 0, and ligation of TEF, and esophago-esphagogostomy were performed on day 10 of life. CoA was corrected on day 15 and at the time, the thymus could not be identified. Hypocalcaemia associated with very low T-cell count and low serum IgG ensued and were suggestive of DiGeorge syndrome. Genetic analysis showed 46, XY without deletion of chromosome 22q11.2. Following complete examination, he was also found to have choanal atresia, abnormal ears, cobolomata, and genital hypoplasia suggestive of CHARGE association. Ca++ and immunoglobulin were administered and the postoperative course was uneventful. He is currently awaiting bone marrow transplantation for treatment of DiGeorge syndrome.

Conclusion: CHARGE and DiGeorge rarely occur together and only 8 cases are reported in the English literature. Two had esophageal atresia and severe congenital heart anomalies. The challenge for management in this complicated case is the background DiGeorge syndrome which can influence surgical outcome because of immune deficiency and Ca++ imbalance which can be life-threatening and require bone marrow transplantation.