A CASE REPORT OF PENTALOGY OF CANTRELL

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We report a case of Indonesian infant boy with a rare Pentalogy of Cantrell which consists of defects of the lower sternum, anterior diaphragm, midline supraumbilical abdominal wall and diaphragmatic pericardium with ectopia cordis. He first presented to HTAR on day 15 of life with supraventricular tachycardia and subsequent admission at 3 months of life with respiratory distress requiring prolonged ventilation. He had an omphalocele at birth and during second admission he was found to have a large central tendon defect of diaphragm with absence of pericardium and intrathoraxic herniation of liver into mediastinum pushing the heart to the left. He had surgical repair of the congenital diaphragmatic hernia, abdominoplasty and right herniotomy done at 5 months of age. The Pentalogy of Cantrell must be actively sought in every patient with an omphalocele.