

CASE 12

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This was the case of a 2-1/2 year old female who was operated upon at six days of age for cyanotic congenital heart disease. The patient was found to have a tricuspid atresia. Definitive therapy consisted of amputation of the pulmonary artery just distal to the pulmonary valve and end to side pulmonary artery-ascending aorta anastomosis. She recovered uneventfully from the surgical procedure and the cyanosis was relieved. The patient also had a right upper lobe pneumonectomy for atelectasis and pneumonia secondary to an anomalous right upper lobe bronchus at the age of 3 months. Following the second procedure, the patient did well until the age of 28 months when she suddenly developed cyanosis and coma. On admission, the limbs were rigid and there was bilateral ankle clonus. She responded only to painful stimuli. A spinal tap revealed clear, colorless fluid under a pressure of 270 mm. The total protein was 7 mgm.%, chloride 130 mEq/L, and sugar 110 mgm.% There were no cells and no growth was reported on routine culture. The child remained comatose and expired on the 23rd hospital day.

Postmortem examination revealed focal destruction of the white matter scattered throughout both cerebral hemispheres. The gray matter appeared not to be grossly involved.