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Unusual Hand Malformation with Cardiac Defect: A Rare Presentation

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ABSTRACT Congenital heart defects occurring together with upper limb malformations as a group of dominantly inherited disorders constitute the Heart-Hand syndrome (HHS) with varied phenotypic manifestations. The researchers report herein a case of HHS Type IV presented with large PDA and cutaneous syndactyly from India. Echocardiography showed large PDA with left to right shunt. Radiograph of the right hands showed features of complete simple cutaneous syndactyly (middle and ring finger) and central type of polydactyly with probable syndactyly (index finger). Karyotype and FISH showed a normal 46,XX. The management of individuals with HHS optimally involves a multidisciplinary team approach, with specialists in genetics, cardiology and orthopaedics including a specialist in hand surgery.