

## VACTERL ASSOCIATION WITH STERNAL CLEFT

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### ABSTRACT

We present a case with Vacterl association. Our patient had imperforated anus associated with cardiac lesion and sternal cleft. Sternal cleft has not been reported before in the literature as a skeletal anomaly with Vacterl association.

**KEY WORDS:** Vactrel, Sternal, Cleft.

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### INTRODUCTION

Anorectal anomalies occur in one of every 599 births and are slightly more common in males.<sup>1</sup> Other associated congenital anomalies can amount to 60%.<sup>2</sup> The incidence of the associated anomalies in a recent study include vertebral anomaly(28%), other skeletal anomaly (13%), cardiac anomaly (10%), tracheoesophagus anomaly(6%), urologic anomaly (31%), vesico-ureteral reflux (2%), spinal cord related anomaly (8%), genital anomaly (8%) and gastrointestinal anomaly (9%).<sup>3</sup> Sternal clefts is a rare anomaly and a recent review of sternal clefts enumerated 73 cases in the literature since 1800.<sup>4</sup> Simple sternal clefts without ectopia cordis are usually without associated malformation.<sup>5</sup> We did not find any listed cases of imperforate anus associated with sternal clefts in the English

language medical literature to date. We present here a case of imperforated anus associated with cardiac lesions and superior sternal cleft.

### CASE REPORT

A 10 hour old male newborn was admitted to our neonatal intensive care unit with imperforated anus and sternal defect. The newborn was delivered by caesarian section at 38 weeks of gestation because of previous caesarian section. Birth weight was 2930g and the APGAR score was 7 and 9 at one and five minute of age respectively. Physical examination revealed superior sternal cleft, midline raphe and absent external anal opening (Figure-1). The heart was lying inside the thorax and was covered by normal skin. The heart was hyper kinetic and no murmur was heard. No fistula was present in the perineum or scrotum. 24 hour after birth high imperforated anus was confirmed radiographically with invertogram which was carried out at the 30th hour after birth.

Chest x-ray, a supine film of the abdomen and pelvic was obtained which did give any evidence of anomalies of vertebra and ribs (Fig-2) Pelvis and abdominal sonography did not define anatomic abnormalities of the upper and lower urinary tract MCUG was not performed. Echocardiography revealed multiple cardiac lesions including pericardial defect, large SD, moderate size VSD and a dilated pulmonary artery. A divided sigmoid colostomy was performed.

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Fig-1: Sternal cleft was seen in the figure.

### DISCUSSION

Imperforated anus results from a failure of differentiation of the urogenital sinus and cloaca. Other associated congenital anomalies occur in more than half of the patients, with genitourinary tract malformation seen in 28%, gastrointestinal in 13%, cardiac in 7%, and skeletal or central nervous system anomalies seen in 6% of affected patients. When these occur in close association, the VACTERL constellation of anomalies should be excluded.<sup>6</sup> Bofto et al reported 286 infants with the VACTERL association, defined as at least three of the five VACTERL anomalies.<sup>7</sup> Seventy percent of babies with VACTERL association have three affected system.<sup>8</sup> Multiple skeletal anomalies other than vertebral anomalies include meromelia of limb,<sup>9</sup> triphalangeal thumb,<sup>10</sup> and left hand rib anomaly. Sternal clefts are rare congenital malformations caused by the failure of fusion of sternal elements during the sixth week of gestation, lateral mesodermal plates move ventrally creating two parallel mesenchymal stripes. The isolated sternal defects are thought to result from the failure of mesenchymal plate fusion process during the eight week of gestation. This failure appears partial or complete. The partial sternal cleft is superiorly or inferiorly located. The inferior type is frequently associated with omphalocele, ectopia cordis, pericardial defect and structural heart defects (Cantrells pentalogy). Sternal clefts associated with gastrochesia were not reported



Fig-2: Anomalies of the vertebra and raphe were seen.

before, to the best of our knowledge. We considered that it is a case of VACTERL association and sternal cleft is a skeletal anomaly which can occur in patient with VACTERL association.

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