microscopic evaluation, has not significantly decreased the incidence of complications following kidney biopsy(4). Hematuria is the most common complication of percutaneous renal biopsy(2). Microscopic hematuria is said to occur in virtually all patients, but gross hematuria has been reported in 2-50% of children biopsied. Significant perinephric hematoma can occur in 0.54-2.9% patients(3-5). Mortality following kidney biopsy is rare but well described. Slotkin and Madsen(3) and Diaz-Buxo and Donadio(4) have reported an overall mortality of only 0.1% in 5000 and 1000 biopsies, respectively.

Pneumothorax following a renal biopsy is uncommon. Slotkin and Madsen found only 9 cases of unilateral pneumothorax in a review of over 5000 cases who had undergone renal, biopsy(3). There is no report of bilateral pneumothorax developing following kidney biopsy. Why our patient developed bilateral pneumothorax is still unclear but the hypothesis is that air can traverse to opposite side through mediastinum in presence of subcutaneous emphysema (which was present in our case) or in presence of mediastinal emphysema, which we could not demonstrate on X-rays. There is increased risk of pneumothorax in young children(4). This may be related to inadequate immobilization of a young child.

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Congenital Complete Heart Block

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Gerbezius in 1718 was the first physician to report the slow pulse of atrioventricular block(l). Morgeni in 1791, published a description of a patient with slow pulse and seizure. However, it was not until 1901 that Morquio described

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several children from the same family with congenital heart block who had slow pulse, syncopal attack and death in childhood(2). Vandee Harvel in 1908, reported ECG findings of complete atrio-ventricular block(3). Considerable information has accumulated since then concerning the clinical profile and circulatory hemodynamics in such cases(4,5). Mortality is highest in neonatal period and major cardiac malformations are a bad prognostic sign(6). We present a case of complete congenital heart block with major cardiac defect in whom an autopsy was performed.

Case Report

A full term newborn female child weighing 3 kg was admitted with history of respiratory distress, tachypnea and groaning since birth. The child was delivered normally at ESI hospital and cried immediately after birth. The antenatal period was uneventful. There was no history of consanguienity, maternal fever, rashes, joint pain or drug intake. The patient was third in order of birth and previous two sibs were normal. There was no history of abortion or still birth.

On examination, the child was hypothermic (temperature 36°C), groaning and cyanosed. Cry, activity and neonatal reflexes were decreased. There was marked respiratory distress; respiratory distress; respiratory rate was 60/min. with intercostal and subcostal recession. Heart rate was 60 per min. All peripheral pulses were palpable and pedal edema was present. Cardiovascular system examination revealed the apex beat on right side in the fourth intercostal space. Heart sounds were normal but better audible on the right side. A pansystolic murmur of Grade III intensity was audible all over precordium. Liver was 2 cm below costal margin on left side. Bilateral crepitations were heard in the chest. No other obvious congenital anomaly was found.-

On investigations blood sugar was 90 mg/dl, oxygen saturation 80% (Hb was 11.75 g/dl, TLC 6800/cu mm and micro ESR: 12 mm). Infantogram was normal and chest X-ray showed dextrocardia with normal sized heart with situs inversus with bilateral infiltration in lungs. On electrocardiogram, atrial rate was 120/min, and ventricular rate was 60/min with complete A-V dissociation. QRS duration was 0.07 sec (normal); QTc was 0.57 sec (prolonged); and few junctional ectopics were seen.

Baby's blood group was B +ve, mother's A +ve and baby had a maximum bilirubin of 12 mg/dl. VDRL of mother and baby both were non reactive and mother's antinuclear antibody test was negative. 2D-Echo revealed complete endocardial cushion defect with left sided ventricle hypertrophy and right side ventricle was rudimentary.

The child was diagnosed as a case of complete congenital heart block with congenital heart disease with situs inversus and chest infection. The baby was managed with nil orally, oxygen inhalation, intravenous fluid, antibiotics and warmer care. Injection atropine in a dosage of 0.03 mg/dose subcutaneously was given whenever required. One blood transfusion was also given. Phototherapy was given for hyperbilirubinemia. Inspite of all resuscitative measures, the child expired of

respiratory failure on 6th day and an autopsy was done within 2 hours.

Autopsy showed dextrocardia with situs inversus and rotation of all organs *(Fig.* 2), a common single atrium with common AV canal opening into both the ventricles. Left ventricle (right sided ventricle) was rudimentary and membranous part of intraventricular septum was absent *(Fig. 2)*. Both the aorta and pulmonary trunk were arising from right venricle (left sided ventricle). Mild pulmonic stenosis was present. Liver and spleen showed severe congestion with foci of extra medullary hematopoiesis. Kidney showed mild cloudy swelling and lungs had bronchopneumonia.

Discussion

The incidence of complete congenital heart block varies from 1:2500 to 1:20000 live births. Occasionally, these cases are symptomatic in neonatal period but whenever they do so they are high risk patients. The mortality is highest during the neonatal period(7,8). In three fourth of the cases of complete heart block, there is pure electrical discontinuity in specialized conducting pathways, whereas in the other group, associated cardiac malformations are found(9). The common anatomical defects mentioned in literature include ventricular septal defect, endocardial cushion defect, patent ductus arteriosus, mitral incompetence, persistent foramen ovale, coarctation of aorta, transposition of grea arteries and Ebstein's anomaly.

Our patient had a common atrium with common atrio-ventricular canal opening into hemodynamically single ventricle. Both the great vessels were arising out of right ventricle. In a study by Robertson of 65 cases of congenital heart block, the highest risk was in patients with anatomic defects, the other risk factors being low ventricular rate (55/min) and prolonged QTc interval in ECG(7). In our child, all the risk factors were present. The low ventricular rate makes the newborn more symptomatic than the older children because of less contractile reserve of heart. At the same time, demand is great due to immense circulatory changes occurring in peripartum period(8). Prolonged QTc is not a consent finding in congenital heart

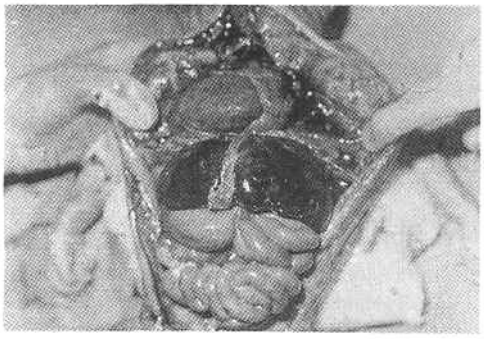

Fig. 1. Dextrocardia with situs inversus totalis.

Fig. 2. Autopsy of heart showing common atrium (CA), Common AV ring (AVR) and membranous ventricular septal defect (VSD).

block but is common with associated malformations and symptomatic patients. In asymptomatic patients, prolonged QTc may herald the onset of symptoms(9). The risk of sudden death is twice great in patients who have a prolonged QTc interval. When all the high risk factors are present in a patient, prognosis remains grave. However, prompt treatment with ionotropic agents, temporary pacing and decongestive measures may save some babies, if diagnosed *in utero*. On the other hand, congenital heart block due to pure electric discontinuity remains asymptomatic and such subjects do fairly well. There is an approximately 95% twenty year survival of patients without anatomic heart defect(7).

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Wiskott-Aldrich Syndrome

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Wiskott-Aldrich syndrome (WAS) is a rare X-linked immunohematological