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The intrathoracic kidney: should we fix it?

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Abstract

Background: Intrathoracic kidney is a rare congenital anomaly, with only 13 cases reported in the pediatric age group over the past 25 years. The relevant literature is limited to individual case reports or small case series with no follow-up data. Both operative and nonoperative management has been advocated. We report our experience in the management of children with an intrathoracic kidney as well as the efficacy of nonoperative management in select patients.

Methods: Five cases of intrathoracic kidney were collected prospectively since 1992 and carefully followed up long term.

Results: Two children presented with acute respiratory distress and underwent right diaphragmatic hernia repair and nephropexy. Incidental diagnosis of a left intrathoracic kidney was made in 3 children. Long-term follow-up has demonstrated normal function and development of these kidneys in all 5 children with no late bowel herniation in the nonoperative group.

Conclusion: Intrathoracic kidney associated with bowel in the chest should undergo standard repair and nephropexy. An isolated intrathoracic kidney without evidence of bowel herniation can safely be observed. This is the largest pediatric series of intrathoracic kidney as well as the first to document the efficacy of nonoperative management with long-term follow-up.

Intrathoracic kidney is a rare congenital anomaly with an autopsy prevalence of 1 in 10,000 [1]. The clinical prevalence is much lower with only 13 pediatric cases reported over the last 25 years [2-9]. The pertinent literature is limited to case reports or very small case series. There is no treatment consensus, and there are very little follow-up data. We describe the 5 cases that we have treated at our institution since 1992 as well as their long-term follow-up.

1. Case 1

A 4-month-old female infant was referred for evaluation of a 24-hour history of progressive respiratory distress. Prenatal ultrasound had demonstrated a single kidney on the left as well as a 2-vessel umbilical cord. Postnatal ultrasound at 1 month of age had confirmed congenital absence of the right kidney, although there was a normally positioned right adrenal gland. The left kidney and adrenal were completely normal. No other congenital anomalies were noted, and the infant did well until acute respiratory problems developed. Chest radiographs showed multiple air-filled bowel loops in the right hemithorax along with pulmonary hypoplasia and some mediastinal shift. An

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abdominal ultrasound showed that the right kidney was displaced superior to a normally positioned liver and just behind the heart. The right adrenal was normally positioned below the diaphragm. At operation, the right kidney was found above the large diaphragmatic defect posterior to the right atrium and just anterior to the inferior cava. It was rotated 90° in a clockwise direction from its normal position. The bowel and kidney were easily mobilized into the abdomen, and primary repair of the hernia and nephropexy were performed. She has been followed up for 14 years with mild hypoplasia of the right kidney.

2. Case 2

A 4-month-old female infant was referred for evaluation of a possible left congenital diaphragmatic hernia found incidentally on a chest radiograph performed for bronchiolitis. The chest radiograph (Fig. 1A and B) showed what appeared to be an intrathoracic kidney (see arrow) with no bowel or stomach in the chest. After recovery from the upper respiratory tract illness, she was completely asymptomatic. Subsequent ultrasounds showed the left kidney in the lower left chest cephalad to the spleen. The kidney was normal in size with no evidence of obstruction. She was followed up with serial follow-up visits, chest radiographs, and renal ultrasounds until she was 17 years old. She never developed any respiratory or gastrointestinal (GI) symptoms. The left kidney developed normally, and there was never any herniation of bowel into the chest.

3. Case 3

An 11-year-old girl presented to the surgery clinic with a history of congenital left intrathoracic kidney diagnosed in India at 4 years old. Apart for rare episodes of mild chest, left flank, and left upper abdominal discomfort, she was completely asymptomatic. Chest radiographs demonstrated a left intrathoracic kidney with no evidence of pulmonary hypoplasia or herniated bowel, stomach, or spleen. On abdominal ultrasound, an intrathoracic left kidney was confirmed. The kidney itself appeared normal in size and parenchymal architecture and was located above the spleen. No hydronephrosis was noted. Because of her vague GI symptoms, both a contrast enema and upper GI series and small bowel follow through were performed. Neither of these studies demonstrated any evidence of bowel or stomach herniation into the chest. Follow-up chest radiographs and renal ultrasounds were performed until she was 17 years old. These demonstrated no herniation of bowel into the chest and normal growth and development of the kidney.

4. Case 4

An 8-month-old female infant was referred to our surgical clinic after a chest radiograph revealed a possible diaphragmatic hernia. The chest radiograph was taken at 5 month of age when she had been hospitalized and treated for 2 weeks for pneumonia. Apart from this hospitalization, the female infant was completely healthy. A chest radiograph repeated at 8 months of age confirmed a left intrathoracic kidney with

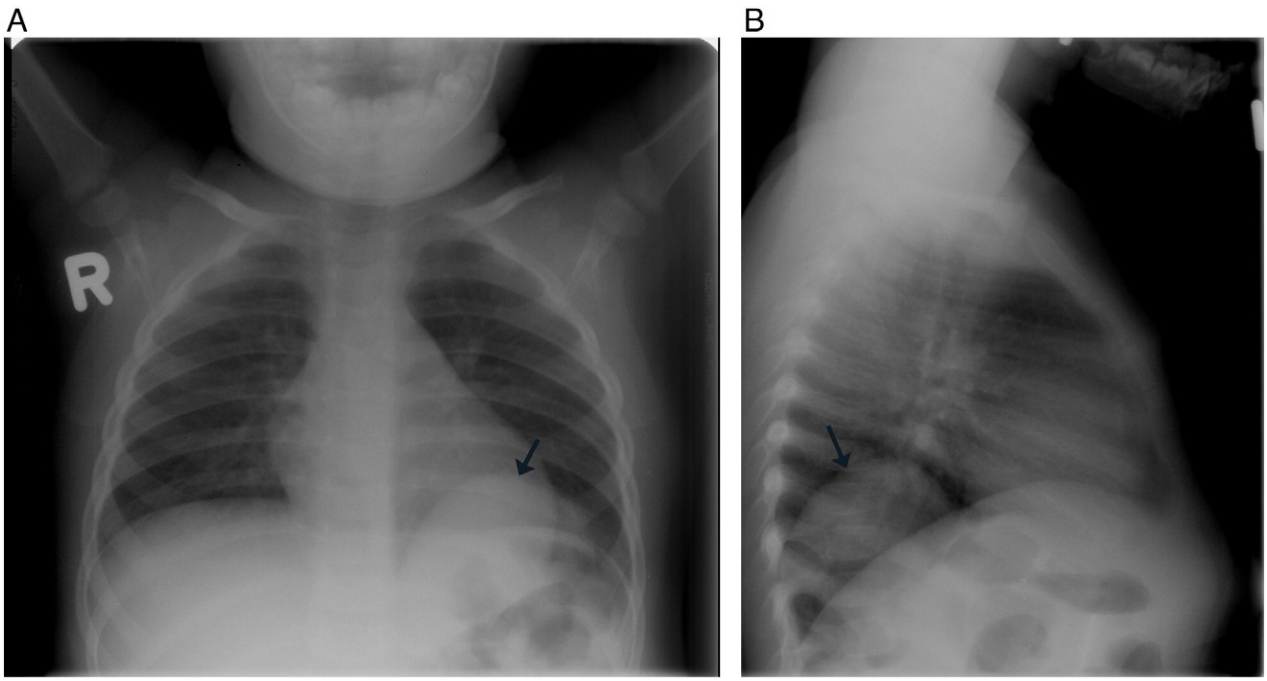

Fig. 1 A, Chest x-ray, posterior-anterior. B, Chest x-ray, lateral.