CONGENITAL SOLITARY FUNCTIONING KIDNEY AND OTHER ASSOCIATED CONGENITAL MALFORMATIONS

Tânia Carvalho¹, Telma Francisco¹, Raquel Santos¹, Margarida Abranches²
¹Department of Paediatrics, Hospital de Ótica da Área de Vila Franca de Xira, Portugal; ²Pediatric Nephrology Unit, Department of Paediatrics Hospital de Dona Estefânia, CHLC – EPE, Lisbon, Portugal

INTRODUCTION AND OBJECTIVE

Congenital solitary functioning kidney (CSFK) is associated to other congenital anomalies. Most of them affect urogenital tract, cardiac, skeletal and central nervous system. There are also some syndromes associated with renal malformations. The objective of this study was to determine prevalence of associated malformations in children with CSFK.

METHODS

We reviewed electronic processes of 134 children with CFSK followed in a tertiary department of paediatric nephrology during five years (2012 – 2016). The congenital malformations found were grouped into system of organs.

RESULTS

<table>
<thead>
<tr>
<th>Urologic</th>
<th>Genital</th>
<th>Gastro-intestinal</th>
<th>Cardio-vascular</th>
<th>Neurological</th>
<th>Otorhinolaryngological</th>
<th>Muscle and skeleton</th>
<th>Ophthalmic</th>
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</thead>
<tbody>
<tr>
<td>Vescoureteral reflux (n=5)</td>
<td>Imperforated hymen (n=1)</td>
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<td>Tetralogy of Fallot (n=2)</td>
<td>Caudal regression syndrome (n=2)</td>
<td>Deafness (n=2)</td>
<td>Vertebral malformation (n=2)</td>
<td>Coloboma</td>
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<td>Pelvic ureteric junction obstruction (n=3)</td>
<td>Uterine didelphys (n=3)</td>
<td>Anorectal malformation (n=4)</td>
<td>Aortic stenosis (n=1)</td>
<td>Spina bifida (n=1)</td>
<td>Inner ear malformation (n=1)</td>
<td>Scoliosis (n=1)</td>
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<tr>
<td>Ectopic ureter (n=1)</td>
<td>Vaginal septum (n=2)</td>
<td>Anorectal atresia (n=1)</td>
<td>Ventricular septal defect (n=1)</td>
<td>Neural geniculocerebellar (n=1)</td>
<td>Cleft palate (n=1)</td>
<td>Abdominal wall defect (n=1)</td>
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<tr>
<td>Ureteroceles (n=1)</td>
<td>Cryptorchidism (n=1)</td>
<td>Ectopic anus (n=1)</td>
<td>Single umbilical artery (n=5)</td>
<td>Dysmorphic cerebellum (n=1)</td>
<td>Limb malformation (n=1)</td>
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<tr>
<td>Obstructive megaureter (n=2)</td>
<td>Hydrospadias (n=2)</td>
<td>Hypospadias (n=2)</td>
<td>Hydrocephalus (n=1)</td>
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</tbody>
</table>

Identified syndromes (n=14)

| VATER/VACTERL (n=2) | Manick Fraser
| Herlyn-Werner-Wunderlich (n=2) | Barakat
| Williams (n=2) | CHARGE
| Mayer Rokitansky Kuster Hauser | Acrorenal mandibular
| Prune Belly | Polymalformative syndrome without specific diagnosis
| Turner |

All identified syndromes were associated with renal anomalies

Majority of cases with renal agenesis had another congenital malformation

Three cases with other congenital malformation had chronic renal disease:

Polymalformative syndrome (1)
Ureteral stenosis (1)
Anorectal malformation (1)

DISCUSSION

This study intends to recall that CSFK may not be the only congenital malformation in a child. It also highlights that there are other anomalies, besides urological tract, that may be present and must be investigated, especially if there is a diagnosis of true renal agenesis. A good prenatal care and careful follow-up of children with CSFK are essential.