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19.1 Introduction

The renal parenchyma is divided into three main compartments:

- The cortex with glomeruli which create the primary ultrafiltrate; the juxtaglomerular apparatus, and the cortical proximal and distal tubules, which are responsible for processing the primary urine thereby maintaining body homeostasis.

- The medulla including the Henle loop performing salt and water reabsorption, involved in concentrating and diluting mechanisms, and medullary collecting ducts for final reabsorption draining the urine through the papilla into the renal collecting system.

- The interstitium, i.e., connective tissue and vasculature, of both compartments (cortex and medulla), including lymphatic tissue.

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Renal parenchymal disease (RPD) is defined as a disease that involves one or more compartments of the renal parenchyma. Although different segments of the nephron, the interstitium, and the vasculature may be affected simultaneously or may become secondarily involved, RPD is generally classified into glomerular, tubular, interstitial, and vascular disease and will be discussed in this order. The causes for similar histological changes and similar imaging appearances of affected kidneys may vary; the same disease can manifest in different ways with a wide range of histology. Furthermore renal parenchyma may be affected by inherited diseases or may become secondarily involved in systemic disease such as metabolic disease, storage disease, infection (e.g., HIV), or sepsis, perfusional disturbances, autoimmune disease, and malignant disease (metastasis, leukemic infiltration, etc.).

Some of the many causes for RPD are discussed and described in other chapters of this book, e.g.: infection and abscess (see Chap. 15), neonatal renal failure (see Chaps. 21, 23), cystic and dysplastic renal disease (see Chap. 10), nephrocalcinosis (see Chap. 20), chronic renal failure (see Chap. 21), changes in congenital urogenital malformations (including obstructive dysplasia, see Chaps. 4–9, 12, 17), traumatic changes (see Chap. 25), or malignant RPD (see Chap. 24). This chapter concentrates on the “classic entities” of RPD such as inherited nephropathy (e.g., Alport syndrome), glomerulonephritis (GN) and nephrotic syndrome (NS), RPD of vascular origin (e.g., haemolytic uraemic syndrome = HUS), and miscellaneous entities such as involvement in metabolic and autoimmune disease.

As well as describing the pathogenesis, main clinical symptoms, histology, and prognosis of the various diseases, this chapter aims to follow the clinical practice in a paediatric radiology department. We list the main findings on initial/primary imaging – generally ultrasound (US) – as established in paediatric radiology (Babcock 1989; Dietrich 1990; Gordon 2003; Hoyer 1996; Kettriz et al. 1996; Mettler and Guiberteau 1998; Riccabona et al. 2001, 2002, 2004, 2005 and 2006; Siegel 1991, 1999; Slovis 1989; Teele and Chare 1991; Toma 1991). Furthermore we try to correlate these US findings with the presenting clinical symptom(s) and the clinical query, and offer important differential diagnoses. We discuss the possibilities and indications for useful additional imaging including renal biopsy, and eventually describe the role of renal and extra-renal imaging during treatment or follow-up of patients with (chronic) RPD.

19.2 Clinical Presentation and Symptomatology

The presentation of renal disease is varied: symptoms may obviously point to the urinary tract, such as macrohaematuria, or—when the kidneys are affected as part of a systemic disease, such as systemic lupus erythematosus (SLE) or metabolic disorders—the initial presentation may be with other systemic manifestation of the disease. Although the aetiology (and the underlying cause, with different treatment and prognosis) may vary widely, the initial presenting features of RPD may be similar. However, serious renal illness may as well be asymptomatic or associated with non-specific symptoms, only coming to light as a consequence of routine examination or screening programs. Laboratory and clinical data can establish the definite diagnosis in only a proportion of cases. Additional imaging often remains inconclusive and history is needed; certain presentations, symptoms and manifestations point to a varying list of differential diagnoses then initiating a specific diagnostic algorithm. The more frequently encountered modes of presentation of RPD are now discussed.

19.2.1 Haematuria

Isolated haematuria is a common finding in childhood and the outcome is benign in the vast majority of cases (Vehaskari et al. 1979). Occasionally, however, isolated haematuria may be an early sign of serious renal disease, e.g., Alport syndrome. Haematuria is classified as either microscopic haematuria with apparently clear urine, or macroscopic haematuria causing a reddish-brown urinary discolouration. The blood may originate from the kidneys or the urinary tract. The microscopic presence of urinary red cell deformity, urinary red cell casts, or proteinuria are characteristic markers of glomerular bleeding (Koehler et al. 1991). Medical history, clinical investigation, and distinction between glomerular and non-glomerular bleeding have a great impact on the diagnostic approach to a child with haematuria. Isolated recurrent or persistent microscopic haematuria is frequently related to idiopathic hypercalciuria, familial essential benign haematuria, minor forms of IgA nephropathy/Henoch-Schönlein nephritis, or acute postinfectious glomerulonephritis.