Nephrotic Syndrome (NS) is a rare diseases (around 2-7 cases per 100.000 children per year) characterized by proteinuria ≥50 mg/kg/day (or ≥40 mg/m2/h) or a proteinuria/creatininuria ratio >2 (mg/mg); hypoalbuminaemia less than 25 g/l and edema. The protein leakage, with the consequent hypoalbunaemia and edema, due to podocyte alterations may be caused by genetic diseases, immunological mechanisms, infections, toxins or malignancy. However, most commonly the exact etiology is unknow. The idiopathic NS may be classified based on response to corticosteroid therapy or the hytological appearance. The first classification identifies steroid-resistant NS (no response after 4 weeks of steroid therapy); frequently relapsing NS (≥ 2 relapses in first 6 months or ≥4 relapses in 1-year); steroid dependent NS (relapses during steroid decalage or within 2 weeks from steroid therapy interruption). The hystological classification is based on light and electron microscopy after renal biopsy, which is indicated in case of onset disease before 1 year or after 12 years of age. Macroscopic hematuria: persistent hypertension and/or microscopic hematuria and/or low plasma C3 renal failure not related to hypovolemia; steroid resistence: secondary or relatedsyndromes NS. Minimal change disease (MCD) is the most common form of idiopahtic NS in children, with good response to steroid treatment, and it is

characterized by normal glomerular appearance on light microscopy and evidence of podocyte foot alterations on electron microscopy, due to immunological related damage. Focal segmental glomerulosclerosis (FSGS) is described inidiopahtic NS, particularly in steroiddependent or steroid-resistant forms, and is characterized by evidence of focal glomerular damage with secondary sclerosis and adhesion with Bowman's capsule; the electron appearance is the same of MCD one. Recent authors hypotizethat the FSGS is an evolution of MCD. These 2 idiopathic NS forms may be expression of the same immunological disease, with 2 different severity grades; so they may be considered different moments of the same disease spectrum. Less common idiopathic NS forms are membrano proliferative glomerulonephritis; membranous nephropathy; IgM-nephropathy; C1q nephropathy and thin basement membrane disease (1, 2, 3).