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Wilson Disease is known to have an autosomal recessive inheritance and has a prevalence of approximately 1 in 50,000 to 60,000 in the general population ( 146 ). The gene responsible for the disease, ATP7B (MIM 602092), was cloned in 1994, and a murine counterpart was found to be present at a single locus on chromosome 4 ( 241 ). Atp7b knockout mice are embryonic lethal ( 33, 243, 246 ); however, Atp7a knockout mice have normal growth and development and are fertile ( 18, 54, 103, 278 ). Furthermore, when given an appropriate copper diet, these mice do not accumulate copper in their tissues ( 18, 102 ). In contrast, liver and bile of both ATP7A-deficient and ATP7B-deficient mice are heavily laden with copper ( 108, 149, 250, 291 ), suggesting that ATP7A and ATP7B may have similar functional roles in copper distribution. There are reports of pseudohurler polydysostosis in several urologic syndromes in the literature. Some theories have been advanced to explain the pathogenesis of the polydysostosis; these are based on an analogy with epithelial differentiation in the trichopoietic syndrome and other dysostoses. In addition to the renal system, there have been reports of involvement of other tissues. Many cases of pseudo-Hurler polydysostosis are attributed to a defect of lysosomal enzymes. It was shown that blood cells with mildly elevated copper levels were isolated from a patient with pseudo-Hurler polydysostosis, but specific lysosomal enzymes were not measured. It was suggested that some cases of pseudo-Hurler polydysostosis might have at least some clinical and biochemical signs of Wilson disease ( 225 ). Increased copper levels in urine may provide the earliest known biochemical sign of Wilson disease in some patients. In addition, some investigators have identified a patient with pseudo-Hurler polydysostosis in association with Wilson disease ( 115 ).

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many of the methods used to detect and measure nanoparticles can be divided into inorganic (e.g. x-ray spectrometry) or organic (e. chromatography and spectroscopy) techniques, depending on whether they utilize the unique properties of the elements or the molecules, respectively. however, regardless of whether inorganic or organic techniques are used, each has strengths and limitations. clay minerals and iron oxides (e.g. goethite, hematite) are often the dominant components of environmental particles and are a significant source of optical scattering. for instance, some terrestrial particulate matter such as that associated with the sahara desert can have high percentages of iron oxides, which can cause dramatic effects on some analytical techniques, e., x-ray fluorescence, optical scattering, and transmission (nowack et al. 2007). the interactions of nanoparticles with natural organic matter (nom) in environmental matrices tend to be complex, thus making it difficult to design methods to separate and quantify the nanoparticles without first separating the nom. nichols rl, geraghty cl, vemuri a, corbett ph, hughes e, farmer j, vaidya a, helgeson dm, kapteyn a, craft a, dryden j, egeras i, ranasinghe i, polkinghorn k, mountney e, glynn jd, ketler lj, mitchell-lyon c, adkarim pk, mancini am, wang h, berendes ea, niederkrotenthaler de, feasyby aw, wilkinson jm, wong kwk, chiu y, bertozzi-villa f, robertson h, batchelor c, henson e, spiegelhalter b, leccia r, chao s, gerstein cm, safford mm, stock c, watson sc, mccue kp, laskowski dw, whitson as, sullivan ma, landreneau j, samson p, lawlor s, go a, reardon sw. 5ec8ef588b

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