Metal mediated bilirubin encephalopathy: Treatment with D-penicillamine

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Very wide-ranging studies have long been made on the possible biochemical transformations of unconjugated bilirubin (UCB) which is formed during the decomposition of hemoglobin. Particular attention has been paid to its photochemical and redox reactions but the relevant publications comprise only a very small proportion of those dealing with the molecular biochemistry of UCB and metal interactions. Bilirubin has a special affinity for the basal ganglia (BG) because they are also target brain regions for divalent metal (Cu, Fe, Zn et cet.) accumulation. The immature and strikingly vulnerable neurons play important role in the pathogenesis of bilirubin induced neurological dysfunction (BIND). Following the developmental period, neurons mature and restrict the apoptotic pathway to permit long-term survival. On the basis of abundant research data and hypotheses, according to our concept, the BIND is a neurodegenerative disease of immature brain caused by accumulation of free metals and UCB-Cu complex (as pro-oxidant) in the BG and other parts of CNS relevant to BIND. During pregnancy, the estrogen levels rise, greatly increasing the retention of copper in the body. This metal will pass through the placenta into the unborn child. So many children are being born with toxic levels of copper and other heavy metals which were stored in the mother’s body. The main comorbidity is the hemolysis. During this process, a great amount of heavy metals may circulate in free form in the bloodstream, and can pass through the blood brain barrier, finding entrance into the CNS as well.

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Spontaneous dissecting distal lenticulostriate artery aneurysm in children

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Aneurysms involving the lenticulostriate artery (LSA) are rare with only few cases reported in children. The authors describe 2 pediatric cases of distal LSA aneurysm in previously healthy children- an 8-year-old girl and a 9-year–old boy, both of whom presented with basal ganglia infarction complicated later with hemorrhage and hydrocephalus requiring external ventricular drain insertion. History and imaging identified dissection as the most likely aetiology. In both cases, conservative management with radiological observation was thought prudent. Follow up imaging at 12-15 weeks demonstrated almost complete resolution of the aneurysm in both cases. The natural history of intracranial arterial dissection remains unclear. There is no consensus on the optimal management; anticoagulation and antiplatelet treatment is contentious. Haemorrhage and death are reported after treatment of intracranial dissection with these agents, which should therefore be used with extreme caution. Both surgical and endovascular treatment have been used to treat LSA aneurysms with satisfactory results but not without potential risks. Treatment decisions are generally dependent on the presentation and location of the dissecting aneurysm as well as individual experience. Conservative treatment with careful clinico-radiological monitoring may be a feasible therapeutic strategy when considering the likely risk and benefits to the patient. The clinico-radiological features, aetiology and complex management of such cases are discussed.

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