Neonatal haemochromatosis risk modification with prenatal administration of gammaglobulin in at risk pregnancies

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Neonatal haemocromatosis is a rare disease characterized by severe hepatic dysfunction associated with extra hepatic siderosis. It is due to materno-fetal alloimmunity with alloantibodies directed against a hepatic antigen that is yet to be defined[1]. The risk of recurrence in following pregnancies once a fetus or newborn is affected is high (60 to 80% of cases). Therapy aiming at diminishing alloimmunity during pregnancy has been suggested to reduce the risk of recurrence and thus, intravenous gammaglobulin from 18 weeks gestational age until term has been recommended[2]. From November 2005 to November 2007, four cases of neonatal haemochromatosis have been diagnosed, treated and confirmed by necropsic examination at Hospital de São João[3]. Two mothers (one of which with a long infertility background) declared the intention to get pregnant again and were referred to prenatal diagnosis where weekly administered intravenous gammaglobulin therapy from the eighteenth week gestation until term was proposed and accepted. Both gestations were uneventful. One of the newborns presented with minor hepatic dysfunction with transient elevated transaminases and bilirubin and increased ferritin and alpha-fetoprotein. The other presented with alloimmune neutropenia (related to the mother's treatment??) and failure to growth, without any evidence of hepatic dysfunction. At six months of age, both babies presented normal liver tests, alpha-fetoprotein and ferritin.

Gestational therapy with intravenous gammaglobulin was associated with favourable outcome with a discrete and reversible alteration in liver function tests in one of the newborns. This therapy should be considered in gestations at risk of neonatal haemochromatosis.