Dubin-johnson syndrome

Dubin–Johnson syndrome is an autosomal recessive disorder characterized by **juvenile conjugated** hyperbilirubinemia. It is caused by a mutation in the gene for the ABCC2/MRP2 pump on chromosome 10q24, which transports glucuronidated bilirubin and some xenobiotics from the hepatocyte to the bile ducts. Conjugated bilirubin is then transported from the hepatocytes into the blood by the ABCC3/MRP3-like pump.

The syndrome was described in 1954. Mild conjugated hyperbilirubinemia develops shortly after birth or in childhood. In addition to jaundice and bilirubinuria, abdominal pain, fatigue, and mild liver enlargement may occur. Symptoms worsen with intercurrent illnesses.

The disease is very rare, but a more frequent occurrence has been described in closed communities (in Iranian Jews it has a frequency of 1 in 1300[1]).

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Content

Links

related articles

- Juvenile hyperbilirubinemia
- Rotor syndrome

Reference

SHANI, M – SELIGSOHN, U – GILON, E. Dubin-Johnson syndrome in Israel. I. Clinical, laboratory, and genetic aspects of 101 cases. *Q J Med* [online]. 1970, vol. 156, p. 549-67, Available from <https://www.ncbi.nlm.nih.gov/pubmed/5532959>. ISSN 0033-5622.

References

- OMIM #237500 (https://omim.org/entry/237500)
- VAN DE STEEG, Evita – STRÁNECKÝ, Viktor – HARTMANNOVÁ, Hana. Complete OATP1B1 and OATP1B3 deficiency causes human Rotor syndrome by interrupting conjugated bilirubin reuptake into the liver. *J Clin Invest* [online]. 2012, vol. 2, p. 519-28, Available from <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3266790/?tool=pubmed>. ISSN 0021-9738 (print), 1558-8238. DOI: 10.1172/JCI59526 (http://dx.doi.org/10.1172%2FJCI59526).

1. SHANI, M – SELIGSOHN, U – GILON, E. Dubin-Johnson syndrome in Israel. I. Clinical, laboratory, and genetic aspects of 101 cases. *Q J Med* [online]. 1970, vol. 156, p. 549-67, Available from <https://www.ncbi.nlm.nih.gov/pubmed/5532959>. ISSN 0033-5622.