

Dubin Johnson Syndrome is a rare genetic liver disorder that tends to affect people of Middle Eastern Jewish heritage disproportionately to other groups. It appears to be associated with clotting factor VII in this population. Symptoms may include a yellowish color to the skin (jaundice), and a liver that is sometimes enlarged and tender. Jaundice, which is caused by excess bilirubin (bile pigment), is usually the only symptom of Dubin Johnson Syndrome. Otherwise a physical examination is normal. The disorder rarely appears before puberty. Occasionally the patient may have an enlarged and tender liver and complain of weakness and a painful abdomen, but the liver will function normally. There may sometimes be a mild recurrence of the jaundice. Pregnancy or use of oral contraceptives may cause the disease to become apparent in women when no symptoms appeared previously. Dubin Johnson Syndrome is inherited as an autosomal recessive genetic trait. Human traits, including the classic genetic diseases, are the product of the interaction of two genes, one received from the father and one from the mother. In recessive disorders, the condition does not appear unless a person inherits the same defective gene for the same trait from each parent. If an individual receives one normal gene and one gene for the disease, the person will be a carrier for the disease, but usually will not show symptoms. The risk of transmitting the disease to the children of a couple, both of whom are carriers for a recessive disorder, is 25 percent. Fifty percent of their children risk being carriers of the disease, but generally will not show symptoms of the disorder. Twenty-five percent of their children may receive both normal genes, one from each parent, and will be genetically normal (for that particular trait). The risk is the same for each pregnancy. Dubin Johnson Syndrome is a rare disease that affects males and females in equal numbers. The disorder can occur in all races. However, among Iranian, Iraqi and Moroccan Jews the incidence is as high as 1 in 1300. In Japan an unusually high incidence of Dubin-Johnson Syndrome was found in an isolated area where there was a high rate of intermarriage. Age at onset can be anytime between 10 weeks of age to 56 years. Treatment of Dubin Johnson Syndrome is symptomatic and supportive. In many cases patients may require no treatment even though they have recurrent mild jaundice. However, metabolism of certain drugs may be affected in patients with Dubin-Johnson Syndrome since many pharmaceutical products are metabolized in the liver. Therefore, medications should be carefully supervised by a physician. Genetic counseling may be of benefit for patients and their families affected by Dubin Johnson Syndrome.