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Dubin-Johnson syndrome

Dubin-Johnson syndrome (DJS) is a disorder passed down through families (inherited). In this condition, you may have mild jaundice throughout life.

Causes

DJS is a very rare genetic disorder. In order to inherit the condition, a child must get a copy of the variant gene from both parents.

The syndrome interferes with the body's ability to move bilirubin through the liver into the bile. When the liver and spleen break down worn out red blood cells, bilirubin is produced. Bilirubin normally moves into the bile, which is produced by the liver. It then flows into the bile ducts, is stored in the gallbladder, and then flows into the digestive system.

When bilirubin is not properly transported into the bile, it builds up in the bloodstream. This causes the skin and the whites of the eyes to turn yellow. This is called jaundice. Severely high levels of bilirubin can damage the brain and other organs.

People with DJS have lifelong mild jaundice that may be made worse by:

- Alcohol
- Birth control pills
- Environmental factors that affect the liver
- Infection
- Pregnancy
- Fasting or dehydration
- Fatigue

Symptoms

Mild jaundice, which may not appear until puberty or adulthood, is most often the only symptom of DJS.

Exams and Tests

The following tests can help diagnose this syndrome:

- Liver biopsy
- Liver enzyme levels (blood test)
- Serum bilirubin
- Urinary coproporphyrin levels, including coproporphyrin I level

Treatment

No specific treatment is required.

Outlook (Prognosis)

The outlook is very positive. DJS generally does not shorten a person's lifespan.

Possible Complications

Complications are unusual, but may include the following:

- Abdominal pain
- Severe jaundice

When to Contact a Medical Professional

Contact your health care provider if any of the following occurs:

- Jaundice is severe
- Jaundice gets worse over time
- You also have abdominal pain or other symptoms (which may be a sign that another disorder is causing the jaundice)

Prevention

If you have a family history of DJS, genetic counseling may be helpful if you plan to have children.

References

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