CONDITION

Dubin-johnson syndrome

Dubin-johnson syndrome is an inherited liver condition that causes occasional jaundice without usually damaging the liver.

WHAT IT IS
Inherited difficulty moving bilirubin from the liver

BODY SYSTEM
Liver and bile system

MAIN SYMPTOMS
Intermittent jaundice, yellow eyes, dark urine

WHAT CAUSES IT
Inherited ABCC2 gene changes

HOW IT IS DIAGNOSED
Blood bilirubin, urine coproporphyrins, sometimes genetic testing

MAIN TREATMENT
Monitoring and trigger management; usually no medicine

CAN IT BE CURED?
No - inherited and lifelong

IS IT AN EMERGENCY?
Sometimes - new jaundice needs assessment

LONG-TERM OUTLOOK
Usually normal health and lifespan

Overview

Dubin-johnson syndrome is a rare inherited condition that affects how the liver handles bilirubin. Bilirubin is a yellow substance produced when the body breaks down old red blood cells.

The liver normally processes bilirubin and moves it into bile, a digestive fluid. In Dubin-johnson syndrome, processed bilirubin does not move out of liver cells as efficiently as it should. Some returns to the bloodstream instead. This is called conjugated hyperbilirubinemia , meaning there is too much processed, or "direct," bilirubin in the blood.

The increased bilirubin can make the skin or whites of the eyes look yellow. This is called jaundice. Urine may also become darker because the processed bilirubin is water-soluble and can pass into urine.

Dubin-johnson syndrome usually does not inflame, scar, or weaken the liver. Most people have no symptoms other than occasional jaundice, and many do not learn that they have the condition until blood tests are performed for another reason. Its main importance is distinguishing it from liver, gallbladder, or bile duct problems that may require treatment.

Symptoms

Symptoms may first appear during adolescence or early adulthood, although the condition can be identified at other ages. Some people never notice any symptoms.

The main features are:

Jaundice may become more noticeable during an infection, physical stress, pregnancy, or after taking certain medicines or hormones. These factors do not cause the condition. They can reveal or temporarily increase the bilirubin problem.

Pale or clay-coloured stools, persistent itching, fever, significant abdominal pain, repeated vomiting, weight loss, or confusion are not typical features of Dubin-johnson syndrome. If these occur, another cause should be considered.

- Yellowing of the eyes or skin that may come and go
- Darker urine during an episode of jaundice
- Blood tests showing increased direct bilirubin
- Occasionally, mild tiredness or abdominal discomfort

Causes and risk factors

Dubin-johnson syndrome is caused by changes in a gene called ABCC2 . This gene provides instructions for making a transport protein called MRP2. The protein helps liver cells move processed bilirubin and some other substances into bile.

When MRP2 does not work normally, processed bilirubin can build up in liver cells and return to the blood. Over time, a dark pigment may collect in the liver, giving it a black or dark brown appearance. Despite this colour change, the liver usually continues to function normally.

The condition follows an autosomal recessive inheritance pattern. This means a person usually has a changed copy of the ABCC2 gene from each biological parent. People with only one changed copy are called carriers and generally do not have the condition.

When both parents are carriers, each pregnancy has:

These chances apply separately to each pregnancy. A genetic counsellor can explain inheritance and testing options in the context of your family.

- A 1 in 4 chance of a child having the condition
- A 1 in 2 chance of a child being a carrier
- A 1 in 4 chance of a child inheriting neither changed copy

Diagnosis

A diagnosis begins with blood tests. These usually show an increased bilirubin level, with most of the increase coming from direct, or conjugated, bilirubin. Other measures of liver health, such as liver enzymes, are usually normal.

Tests may also be used to rule out more common reasons for jaundice. These can include hepatitis, blocked bile ducts, gallstones, medicine-related liver injury, and conditions that cause unusually rapid breakdown of red blood cells.

A urine test for substances called coproporphyrins can strongly support the diagnosis. In Dubin-johnson syndrome, the total amount is often normal, but more than 80% is usually coproporphyrin type I. This pattern differs from that seen in people without the condition and from the pattern in Rotor syndrome, another inherited cause of direct hyperbilirubinemia.

Genetic testing can identify changes in both copies of the ABCC2 gene and confirm the diagnosis. It may be especially useful when laboratory results are unclear or when testing could help relatives understand their risk.

An ultrasound or other scan may be performed when a blockage or structural liver problem needs to be excluded. Imaging does not confirm Dubin-johnson syndrome.

A liver biopsy is rarely necessary. If performed for another reason, it may show coarse, dark pigment inside liver cells. Because blood, urine, and genetic tests are usually enough, an invasive biopsy is generally avoidable.

Treatment and management

Dubin-johnson syndrome usually needs no specific medical treatment. There is no standard medicine that corrects the inherited transport problem, and treatment to lower bilirubin is not usually needed because the condition does not generally damage the liver.

Management focuses on confirming the diagnosis and avoiding unnecessary investigations or treatment. Keeping a record of the diagnosis can be helpful if jaundice is noticed during future

medical care.

Illness, dehydration, pregnancy, hormonal contraception, and some medicines may make jaundice more visible in some people. Do not stop a prescribed medicine or contraception on your own. If jaundice begins or becomes bothersome after starting something new, the prescriber can review whether it may be contributing and whether an alternative is appropriate.

The MRP2 protein also helps transport some medicines and their breakdown products. The clinical effect of ABCC2 changes is not fully predictable and is not the same for every medicine. Tell healthcare professionals and pharmacists that you have Dubin-johnson syndrome when medicines are being reviewed.

During pregnancy, bilirubin may rise and jaundice may become more noticeable. The condition itself usually does not prevent a healthy pregnancy, but new or worsening jaundice still needs assessment because pregnancy can also cause unrelated liver and bile conditions.

Complications

Dubin-johnson syndrome is considered benign, meaning it usually does not lead to progressive liver damage. It is not expected to cause cirrhosis, liver failure, or a shortened lifespan.

The most common practical problem is that recurrent jaundice can be mistaken for hepatitis, a blocked bile duct, or another liver condition. This may lead to worry, repeated tests, or procedures if the established diagnosis is not known.

A person with Dubin-johnson syndrome can still develop unrelated liver, gallbladder, or blood conditions. A previous diagnosis should not be used to explain every future episode automatically, particularly if the symptoms or blood-test pattern have changed.

Visible jaundice can also affect confidence or cause concern at work and in social settings. A written explanation that the condition is inherited, non-infectious, and usually harmless may help.

Prevention

Dubin-johnson syndrome cannot be prevented because it is inherited. Diet, exercise, alcohol avoidance, or supplements cannot change the ABCC2 gene or cure the condition.

If the condition runs in your family, genetic counselling can explain carrier testing, inheritance, and reproductive testing options. Whether to have genetic testing is a personal choice.

Although the condition itself cannot be prevented, unnecessary episodes of more noticeable jaundice may sometimes be reduced by staying adequately hydrated and obtaining care for infections. Avoiding all medicines or hormones is neither necessary nor appropriate. Any possible trigger should be reviewed individually rather than stopped without advice.

Living with it and what to expect

Dubin-johnson syndrome remains present throughout life because the underlying gene changes do not go away. However, bilirubin levels can rise and fall. Jaundice may appear for several days or longer and then fade, or it may remain mildly visible.

Most people continue their usual education, work, exercise, travel, relationships, and family life. The condition generally does not require a special diet or routine restriction of physical activity. Regular liver treatment is not usually needed once the diagnosis is secure.

Some people have occasional blood tests, particularly if symptoms change or medicines are being reviewed. Routine follow-up may be limited when bilirubin is stable, other liver tests are normal, and there are no new symptoms. The appropriate schedule is based on the person's wider health and the certainty of the diagnosis.

Over months and years, the condition is not expected to progress into liver failure. Episodes may still become more visible during illness, pregnancy, or other physical stress. A different pattern-such as steadily rising bilirubin, abnormal liver enzymes, pale stools, pain, or itching-needs reassessment rather than being assumed to be part of Dubin-johnson syndrome.

When to get medical help

Arrange a routine medical appointment if:

Seek urgent medical assessment if jaundice occurs with fever, persistent vomiting, marked itching, pale stools, worsening abdominal pain, or unusual weakness. These are not typical of uncomplicated Dubin-johnson syndrome and may indicate a different liver, gallbladder, bile duct, or blood problem.

Seek emergency help for severe abdominal pain, confusion, fainting, difficulty waking, vomiting blood, black stools, or signs of a serious allergic reaction. Dubin-johnson syndrome itself does not usually cause these symptoms.

- You develop yellow eyes or skin for the first time

- Previously diagnosed jaundice becomes more frequent or persistent
- A new medicine seems to make jaundice worse
- You are pregnant and notice new or increasing jaundice
- You want information about genetic testing or family planning

Common questions

Is Dubin-johnson syndrome dangerous?

It is usually not dangerous. It can cause recurrent jaundice, but it does not generally inflame or scar the liver. Life expectancy is usually normal. New or unusual symptoms still need assessment because having this condition does not prevent other health problems.

Will the jaundice go away?

An episode may fade as the bilirubin level falls, but future episodes can occur. Some people have only mild changes that are noticeable mainly on blood tests. Others develop visible yellowing during illness, pregnancy, dehydration, or other physical stress.

Can I pass it to my children?

Yes, the ABCC2 gene changes can be passed on. A person with Dubin-johnson syndrome usually passes one changed copy to each child. Whether a child has the condition also depends on the other biological parent's genes. A genetic counsellor can explain the possibilities for a particular family.

Do I need a special diet?

No special diet has been shown to correct the bilirubin transport problem. A balanced diet and adequate fluid intake support general health, but they do not cure the condition. Products marketed as liver cleanses or detox treatments are not necessary and may contain harmful ingredients.

Can I drink alcohol?

Dubin-johnson syndrome itself does not create a specific alcohol treatment requirement. Alcohol can still affect the liver for other reasons. If you have another liver condition, take medicines that interact with alcohol, are pregnant, or have been advised not to drink, follow that advice.

Is it the same as Rotor syndrome?

No. Both are inherited conditions that cause increased direct bilirubin without usually damaging the liver. They involve different transport proteins and have different urine

coproporphyrin patterns. Dubin-johnson syndrome also causes dark pigment to collect in the liver, while Rotor syndrome does not.

Sources

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