Caroli disease is a rare inherited disorder characterized by abnormal widening (dilatation) of the ducts that carry bile from the liver (intrahepatic bile ducts). According to the medical literature, there are two forms of Caroli disease. In most cases, the isolated or simple form is characterized by widening of the bile ducts (dilatation or ectasia). A second, more complex form is often called Caroli syndrome. The complex form or syndrome is associated with the presence of bands of fibrous tissue in the liver (congenital hepatic fibrosis) and high blood pressure in the portal artery (portal hypertension. This form of Caroli disease is also often associated, in ways that are not well understood, with polycystic kidney disease, and, in severe cases, liver failure. The simple form, Caroli disease, is apparently much less common than is the more complex form. Both forms are more common in females than among males. Most often the disorder presents in adults, although cases of neonatal and childhood presentation of symptoms are recorded. When the liver and spleen are unusually large (hepatomegaly and splenomegaly) and intermittent stomach pain is present, the doctor may ask for imaging studies (such as ultrasound and CT scans) to be done. The results of these studies may lead to a diagnosis of Caroli disease.