My Baby Has a Positive Newborn Screening Result:  
Short Chain Aacyl-CoA Dehydrogenase Deficiency

What is newborn screening?  
These are routine tests done shortly after birth on every baby born in Manitoba. A small sample of blood is taken from your baby and is tested for rare, treatable diseases, including short chain acyl-coA dehydrogenase deficiency (SCADD).

What does it mean if my baby has a positive SCADD newborn screening result?  
This result does not mean that your baby has SCADD. However it means that more testing is needed because of the very small chance that your baby might have a serious form of SCADD that will need treatment (most forms of SCADD do not). In any case babies with SCADD can grow and develop normally if SCADD is diagnosed early, so it is important to have follow-up testing done quickly to find out if your baby has SCADD and whether it is a form that needs treatment.

You may feel worried about your baby's screen positive result. Many parents in this situation feel this way. Remember, we do not know for sure that your baby has an important form of SCADD until more testing has been done.

What is SCADD?  
SCADD is an inherited (genetic) disease that can cause a baby to have problems using fat as an energy source. The human body needs energy to perform all of its daily activities. The body’s main source of energy is a type of sugar called glucose. If a baby has not eaten for a while – such as when they are sick or if they miss a meal – the body has used up all its glucose stores and switches to using fat as an energy source.

The vast majority of babies with SCADD have a “biochemical variant” that is identified on newborn screening but does not cause any problems to your baby. On rare occasion when a baby has an important form of SCADD, his/her body may not be able to switch to using fat for energy. Some babies with SCADD can get very sick if their bodies are not able to use to use fat for energy when needed. However, most babies with SCADD never develop any health problems associated with SCADD; however, there is no way to know who will develop problems caused by SCADD and who will not.

SCADD was originally thought to be very rare. However newborn screening for this disorder revealed that SCADD is more common than previously believed because most have a very mild variant of this condition.

What causes SCADD?  
SCADD happens when an enzyme called short chain acyl-co-A dehydrogenase is either missing or not working properly. This enzyme’s job is to break down the “short” sized fats that we eat, or that are stored in the body, into smaller sized fats and eventually into energy.
How do I find out if my baby has SCADD?
Blood and urine tests are done to find out if a baby who screened positive actually has SCADD.

When can my baby have these tests?
Your baby’s doctor or a health care professional at the Winnipeg Children’s Hospital will call you to talk about the results of your baby’s positive newborn screen and arrange more testing as soon as possible.

Why screen for SCADD?
SCADD is a highly variable and not well understood. Although we have said that most babies found to have SCADD through newborn screening will never have symptoms it is important to know for sure and be prepared. Things that cause stress, such as lack of sleep, going without food for too long, illness, or infection are thought to trigger episodes of illness called metabolic crisis in some SCADD children but not others. A metabolic crisis is a serious health condition caused by low blood sugar and/or the build-up of harmful substances in the blood. Symptoms of a metabolic crisis are poor feeding, vomiting, lethargy, excessive sleepiness and irritability. The goal of screening for SCADD is to identify a baby at birth with a serious form before symptoms appear and with early treatment prevent a metabolic crisis from occurring.

How is SCADD treated?
Babies with SCADD are treated and monitored by a team of specialists including a metabolic doctor, metabolic genetic counsellor, and a dietician. Treatment is recommended for some, but not all, children with SCADD. This may include: frequent feeding and avoiding fasting fasting (in infancy this general means 6 to 8 hours). Sometimes, carnitine or cornstarch is given orally. With early diagnosis and careful management, children with SCADD can live healthy lives with normal growth and development.

How does a baby get SCADD?
SCADD is an inherited (genetic) disease. A baby with SCADD inherited two non-working copies of the SCADD gene, one from each parent. People who have one non-working copy of the SCADD gene are called “carriers.” Carriers of SCADD are healthy and do not have symptoms of SCADD.

Where can I get more information?
For more information on newborn screening, please talk to your local health care provider or visit the Winnipeg Metabolic Program website www.wrha.mb.ca/prog/genetics or the newborn screening section of the Cadham Provincial Laboratory website www.gov.mb.ca/health/publichealth/cpl/baby.html.

For more information on SCADD, please visit the Fatty Acid Oxidation Family Support Group website at http://www.fodsupport.org

NOTE TO PARENTS/GUARDIANS: This information is only for parents whose baby has had a positive newborn screening result for short chain acyl-co-A dehydrogenase deficiency (SCADD). Please remember that this fact sheet was written for information purposes only. The fact sheet should not replace professional medical advice, diagnosis or treatment.