Fatty Acid Oxidation Defects
And Exercise

Why do I experience pain on exercise?
People who have fatty acid oxidation defects may experience pain and/or weakness when they exercise. This is because they have a fault somewhere in the mechanism that breaks up the 'fuels' in our food and converts them into energy. They experience pain when they exercise because the muscles are placing extra demands on the mechanism that breaks up 'fuels'. Because there is a fault with the mechanism, it cannot cope and breaks down.

What food do our muscles need to allow us to exercise?
We all consume carbohydrates (sugar and starches) and fat in our diet. Some of the carbohydrates and fat are used immediately for energy; most is stored for later use. These stored 'fuels' are broken down in the body to provide energy when we need it. This energy is used in all organs; for example in the heart to allow the heart to pump blood and in the brain to allow us to think and control our bodies. Muscles also use a great deal of this energy to allow us to exercise. When we are at rest, our energy use is quite low, but when we exercise the muscles demand a great quantity of energy very quickly. Which fuel is used, carbohydrate or fat, is dependent upon the type and duration of the exercise is carried out. During short duration exercise of high intensity, for example sprinting and weight lifting, the main source of energy comes from carbohydrates. If the exercise is more prolonged, for instance a three-kilometre jog, biking or a game of football, there is an initial use of carbohydrate with a gradual switch to fat as a major provider of energy. This occurs because most of the carbohydrate reserves become depleted and the muscles have to start using stored fat to provide the energy.

How are carbohydrates and fat converted into the energy we need to exercise?
Carbohydrates and fat are stored in different ways, and are broken down to provide energy by different processes. Carbohydrates are broken down into glucose that is then stored as glycogen. Glycogen is stored in the muscles, heart, liver and brain. When we exercise intensely in short bursts, the glycogen is converted back into glucose and then into a substance called pyruvate. The pyruvate then enters the mitochondria (the 'powerhouses' in cells whose major function is to produce energy units known as ATP). If the exercise is strenuous it can overload the system which cannot function fast enough to convert all the pyruvate into energy via the mitochondria. As a result, some of the pyruvate is
Fat is more of a storage fuel. It is stored in various places such as under the skin. We usually use fat for energy during prolonged exercise. The fat is brought out of the tissue where it is stored, goes into the bloodstream and then into muscle, and finally continues into the mitochondria. Once inside the mitochondria, the fats are broken down into carbon dioxide, water and energy (ATP) during a process called fatty acid oxidation.

How do fatty acid oxidation disorders fit in?
People who have these disorders are unable to process fat into energy. The breakdown occurs at the point of fatty acid oxidation.

What type of exercise brings on symptoms?
Symptoms generally occur during prolonged exercise, such as going for a long run, biking or playing a full game of football. The symptoms vary from person to person. The important thing is to find your own limits and stay within them. Sometimes you will experience symptoms when you are feeling unwell or fasting. Fasting is another occasion when we use up the body’s fat stores to provide energy.

What are the symptoms and what should I do if I experience them?
You may feel muscle pain (either an ache or very intense pain) both during and after exercise. The pain can even come on a couple of hours after you stop exercising. When you feel pain, you should immediately stop exercising and should try to avoid continuing that form of exercise for such a long period in the future. Rhabdomyolysis can also occur during prolonged exercise. This occurs when muscle begins to break down and releases myoglobin into the urine. This causes your urine to become noticeably darker than usual (a brownish colour). This is a very serious side effect as it may lead to kidney failure. If you experience rhabdomyolysis please stop the exercise that you are doing and go to the nearest emergency room.

Diet is extremely important in alleviating the symptoms of these conditions. You should see a metabolic dietitian and follow their expert advice.

Frequent glucose drinks should be taken to try and ensure that you never entirely rely on fat for energy. The effects of fatty acid oxidation disorders can be potentially serious. They can lead to severe muscle weakness that can lead to life threatening complications. Restricting prolonged exercise and diet intervention can be an effective method of treatment and prevention.

Adapted from: Turnbull. Metabolic disorders that cause pain and/or weakness on exercise. www.muscular-dystrophy.org