

# BECKWITH-WIEDEMANN SYNDROME (BWS)

## HYPOGLYCEMIA DIAGNOSIS AND MANAGEMENT

**Beckwith-Wiedemann syndrome (BWS)** is a rare disorder involving changes on a region of chromosome 11p15 that influence pre- and postnatal growth. These changes disrupt the normal balance of growth gene expression and lead to the overgrowth seen in patients with BWS. Depending on which parts of the body are affected, children with BWS can have different features. These include overgrowth of the pancreas leading to hypoglycemia or hyperinsulinism.

### What is hypoglycemia?

**Hypoglycemia** is low blood sugar. It is important for the body to have normal blood sugar levels because low sugar levels can cause problems including seizures and brain damage. Approximately half of infants with BWS will have hypoglycemia. In most of these children, hypoglycemia will only last for a few days and can easily be treated with frequent feedings and/or medical doses of sugar (dextrose infusions). However, in 5-10% of children with BWS, hypoglycemia can persist, requiring additional monitoring and medical care.

### What causes hypoglycemia in children with BWS?

Although hypoglycemia can happen for many reasons, in children with BWS, it is usually caused when the pancreas makes too much insulin (**hyperinsulinism**). **Insulin** lowers blood sugar (also called glucose) levels. We suspect that low blood sugar levels in BWS have to do with the genes on chromosome 11p15 that control growth. Additionally, there are some other genes on chromosome 11 that affect how some of the cells in the pancreas work.

### How do we test for hypoglycemia?

A normal blood sugar level is 70 – 120 mg/dL. Low glucose levels are common in the first 24 hours of life; after the second day of life, glucose levels should normalize. A baby with hypoglycemia persisting beyond the first two days of life requires evaluation. This evaluation consists of a **diagnostic fast** to measure insulin levels and other markers. Diagnostic fasts should be monitored by physicians familiar with hyperinsulinism.

### How do we manage hypoglycemia in children with BWS?

Treatment for hypoglycemia is currently based on the clinical needs of each patient. For newborns with hypoglycemia lasting more than a week, frequent feeding and intravenous or oral sugar, such as **dextrose**, can be given to help increase blood sugar levels. Medications like **diazoxide** and **somatostatin analogs**, which reduce the amount of insulin released by the pancreas, can also help.

If hypoglycemia persists despite these efforts, surgery removing part of the pancreas (partial pancreatectomy) may be needed. By removing some of the cells that produce insulin, this surgery can help lower insulin levels and allow blood sugar levels to return to normal. More detailed information is available through the Hyperinsulinism Center at CHOP.

### Summary

- In BWS, half of newborns have low blood sugar levels due to high insulin levels. Hypoglycemia persists in 5 – 10% of these infants.
- Persistent low blood sugar levels can be diagnosed with a diagnostic fast.

- Milder cases of hypoglycemia can be treated with medical doses of sugar and medications that reduce insulin levels.
- In severe cases, surgical removal of the part of the pancreas may be necessary.

Patient family education materials provide educational information to help individuals and families. You should not rely on this information as professional medical advice or to replace any relationship with your physician or healthcare provider.