Hyperglycemia is a common event in neonates, frequently associated with specific clinical conditions (sepsis, drugs or intravenous fluids) other than neonatal diabetes. Unusual endocrinometabolic syndromes must be considered whenever initial studies are inconclusive.

**NEWBORN NICU ADMISSION**
- Low birth weight
- Hypotonia
- Feeding difficulties

**PRENATAL HISTORY**
- Healthy parents
- IG IP: 37WG Late IUGR
- Oligoamnios / Fluxometric changes
- Maternal hypertension (35 W) and autoimmune thyroiditis
- Eutocic delivery
- Apgar 8/10
- W: 1710 g (<<10)
- H: 43 cm (<<P10)
- HC: 29 cm (<<10)

**DAY11**
- Tachycardia
- Hyperglycemia
  - 180 - 349 mg/dL
- CBC, venous blood gas
- Blood cultures
- Ketonemia/Ketonuria
- Negative

**DAY 28**
- Tachycardia
- Progressive hypotonia
- Hyperglycemia
  - 200 mg/dL
- Hypertrygliceridemia
  - 3549mg/dL

**DAY 34**
- Discharged home
- TG levels - 175mg/dL
- Net weight increase
  - 20g/day

**FOLLOW UP**
- Lipodystrophic phenotype gradually developed
- Insulin levels persistently elevated (no hyperglycemia)
- Hepatomegaly and steatohepatitis
- TG 400-800mg/dL

**BSCL2 gene sequenced:** Compound Heterozygous (c.399C>A (p.Tyr133*) and c.604C>T (p.Arg202*))

**BERARDINELLI-SEIP SYNDROME**

**At 15 months**
- Borderline inferior psychomotor development
- Breast button without precocious puberty and neck acanthosis nigricans
- Normal growth velocity, and concordant skeletal age
- Concentric hypertrophic cardiomyopathy (IV septum 9mm)
- Under diet with 11% lipid, 15% protein and 74% carbohydrates + lcPUFAs

The authors present a case of Berardinelli – Seip Syndrome (Congenital generalized lipodystrophy type 2) as the primary cause of neonatal insulin resistance and hyperglycemia. This syndrome is characterized by the absence of functional adipocytes with storage of lipids in muscle and liver and consequent hepatomegaly, steatosis, and skeletal muscle hyperthrophy. Besides early insulin resistance, other endocrine manifestations that should be regularly surveilled include diabetes mellitus of difficult control, hirsutism, and precocous puberty.