First Nations Health Authority
Health through wellness

Preventing and Managing Hypoglycaemia in First Nations Infants and Young Children in BC

PSBC: Healthy Mothers, Healthy Infants, 2016

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Learning objectives

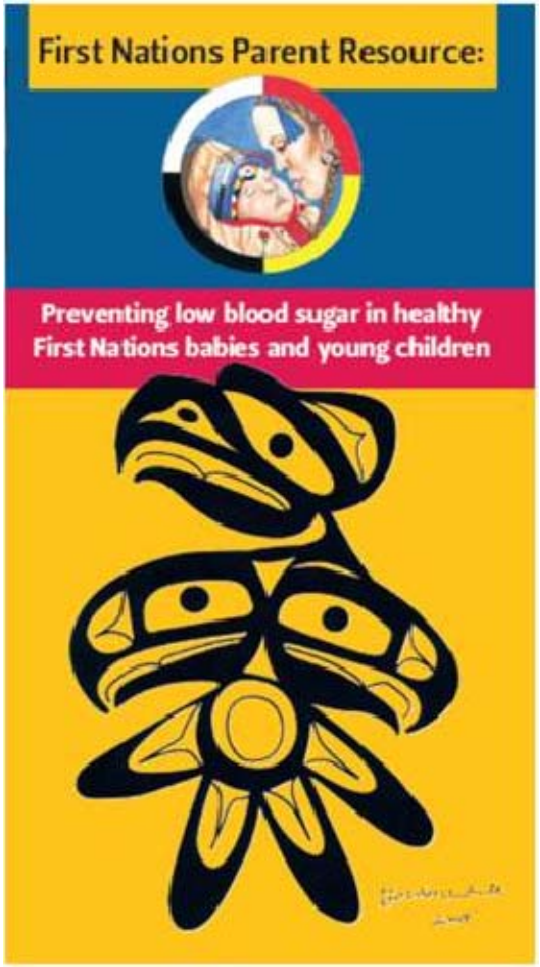
1. Describe CPT1a
2. Identify the significance of the disorder
3. Identify the health promotion and wellness strategies related to CPT1a
4. Describe the available resources
5. Understand the importance of the key take home messages

Case study

Background

- Some First Nations infants and young children may be at increased risk of hypoglycaemia (low blood sugar)
- This increased risk is due to a common genetic variant in the CPT1a gene, which results in a slower rate of fatty acid oxidation
- Very low blood sugar can cause brain injury

Tripartite working group


First Nations Parent Resource:
Preventing low blood sugar in healthy First Nations babies and young children

Medical Guideline: Prevention and Management of Hypoglycaemia in First Nation Infants and Young Children Including Screening for CPT1a Variant in Infants and Young Children who Present with Ketotic and Hypoketotic Hypoglycemia

If you need information or advice on the management of an infant or child with CPT1a variant, please do not hesitate to call the Biochemical Disease physician on call through BC Children's Hospital paging service at (604) 875-2161.

Biochemical Disease Physician On Call: 604-875-2161

PREVENTION AND MANAGEMENT OF HYPOGLYCAEMIA IN BC FIRST NATIONS' INFANTS AND YOUNG CHILDREN INCLUDING SCREENING FOR CPT1A VARIANT IN INFANTS AND YOUNG CHILDREN WHO PRESENT WITH KETOTIC AND HYPOKETOTIC HYPOGLYCEMIA

AUTHORS:

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- Lucy Barney, Provincial Lead, Aboriginal Health, Perinatal Services BC
- Charmaine Enns, Medical Health Officer, Island Health
- Melanie Foster, Policy Analyst, Child and Youth Health, Ministry of Health
- Carolyn Henson, Director, Child and Youth Health, Ministry of Health
- Graham Sinclair, Biochemical Geneticist, BC Newborn Screening Program, BC Children's Hospital
- Sylvia Stockler, Program Director, Biochemical Diseases, BC Children's Hospital
- Hilary Vallance, Medical Director, BC Newborn Screening Program, BC Children's Hospital
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Last updated: October 9, 2014
Lead for medical guideline revisions: Dr Hilary Vallance

1.0 BACKGROUND

The development of this medical guideline was initiated in relation to the observation that some BC First Nations' infants and children may be susceptible to hypoglycaemia during prolonged fasting and/or illnesses that could interfere with feeding due to a common genetic variant and that

What is CPT1a?

- Carnitine palmitoyltransferase 1 (CPT1a) key regulatory enzyme for the oxidation of fatty acids,.
- During a fasting state, CPT1a enzyme is activated and fatty acids are broken down to produce energy and ketones.
- Inherited condition 2 forms: CPT1a deficiency (rare autosomal recessive disorder) or CPT1a variant
- CPT1a variant common genetic variant: results in partial loss of enzyme activity – slower rate of fatty acid oxidation
- Both parents must have the variant for an infant to have the variant

CPT1a continued

- First Nations babies born with the CPT1a variant:
  - Coastal BC and Vancouver Island – 1 in 5
  - Interior BC – 1 in 25

- No universal screening for the CPT1a variant
  - vast majority of people born with it have no health issues

- Important to not cause undue alarm amongst parents, as CPT1a is common and screening is not the answer – health promoting messaging about feeding is key

Health promotion is the best medicine:

- Promote and support breastfeeding
- Ensure if not breastfeeding the use of appropriate formula
- Feed baby on demand and ensure frequency of feeding = age of baby/child
- Offer breastmilk (formula/other food) before bed and after waking in am
- Ensure parents know normal sleep patterns
- Promote safe sleeping

Health promotion

Educate nurses, doctors, parents and care providers:

- When baby/child appears mildly sick:
  - -increase frequency of feeds
  - -wake up child 1-2 x during the night
- When to seek medical attention:
  - -fever, vomiting, diarrhea
  - -refuses feeds
  - -excessively sleepy and hard to wake up
  - -decreased urine and stool for age
  - -signs dehydration (parents know them)

Challenges

Acute care management

- Know your client
- Check blood sugar
- Test for ketones in urine
- Rule out other causes of hypoglycemia
- Consider CPT1a variant and testing
- Treat hypoglycemia

Child with CPT1a

- Healthy babies and children usually do not have problems
- Health promotion strategies
- Special consideration for situations where fasting required
- Consult pediatrician for dx or continued care
- Ensure parents, family, caregivers are well informed
- Provide parent brochure
- Parents have letter of explanation

Resources

CHILD HEALTH BC
LEAD BENEFACTOR
save on foods

http://childhealthbc.ca/

Guidelines, Reports, and Presentations

Files / Hypoglycemia in BC First Nations Infants and Young Children (CPT1a) /

- pdf Medical Guideline
- pdf Parent Resource

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Key take home messages

- Increase awareness with health care providers
- Access resources: Child Health BC website
- Listen to parents/family/foster parents
- Consider CPT1a during assessment / dx
- Test blood sugars
- Consider IV for baby/child fasting >4hrs

Key take home messages continued

- Strong message to feed baby frequently
- Ensure everyone caring for baby is educated
- Ensure families know when to seek medical attention when baby is sick.
- Reassure parents under normal circumstances there will be no problems,
- Children outgrow and are fine

Moving forward

- Continue education: webinars, UBC learning circle, conferences, education forums
- Spread the message to all health care professionals
- Ensure people are aware of resources
- Ensure website is up to date.

Healthy Outcomes

Thank you to our Working Group

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Questions?

Comments?

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