Nutrition Interventions for ROHHAD

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Advanced Pediatric Case Study

Outline

- Discuss pathophysiology and clinical presentation of ROHHAD.
- Introduce case study patient.
- Detail nutrition interventions taken as part of medical management of ROHHAD.

What is ROHHAD?

Rapid-onset obesity with hypothalamic dysfunction, hypoventilation, and autonomic dysregulation


Hypothalamic Dysfunction

Disease

Hypothalamus

Body temperature, growth, salt and water balance, sleep, weight, appetite, and more

Hypothalamus detects changes in

Pituitary Gland releases hormones to

Adrenal glands, thyroid gland, ovaries, and testes

Hypothalamic Dysfunction

- Water imbalance
  - Dysregulation of antidiuretic hormone (ADH)
  - Hypernatremia or hyponatremia
- Hypothyroidism
- Growth hormone deficiency
- Altered onset of puberty

Hypothalamus

**Hypoventilation**

- Develops either with or after onset of obesity.
- Consists of obstructive sleep apnea and/or central hypoventilation.

**Autonomic Dysregulation**

- **Autonomic Nervous System**
  - Receives input in the body and regulates physiologic processes.
  - Controls BP, HR, body temp, weight, digestion, metabolism, fluid and electrolyte balance, sweating, urination, and more.

- **ROHHAD Symptoms**
  - Decreased heart rate
  - Ophthalmologic abnormalities
  - Altered thermoregulation
  - Gastrointestinal dysmotility
  - Altered pain perception

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**Case Study Patient**

- 16 y/o Caucasian female
- Resident at skilled nursing facility for 10 years, multiple discharges and readmissions for medical procedures
- Attended school at out-of-house facility
- In custody of the Division of Child and Family Services
- Parents have visitation rights

**ROHHAD Diagnosis**

- Initial admit at skilled nursing facility (age 6)
  - Admit dx: excessive weight gain of unknown etiology (similar to Prader-Willi syndrome), obstructive sleep apnea, severe dependent at night
  - Nutrition note: “Resident at severe nutritional risk w/ excessive weight gain of unknown etiology, and seemingly unmeasurable appetite. Resident is at 200% IDEW with a BMI of 32. Physician stated in pediatric rounds that it is thought that weight gain may be a central endocrine abnormality but is yet to be diagnosed…”

- 8/30: Discharged to acute care hospital for hypoventilation, bradycardia, and low body temperature (BP 77/47, pulse 44, temp 35.8, RR 16). Diagnosed with ROHHAD. Pacemaker placed.
- 9/30: Readmitted to skilled nursing facility.

**Medical History**

- Central hypoventilation with respiratory failure and trach-vent dependence
- Pituitary insufficiency with thyroid replacement and growth hormone
- Behavioral problems
- Developmental delay
- Obstructive sleep apnea
- Chronic lung disease with pulmonary fibrosis
- Central temperature instability
Medical Treatment

- Rapid Onset Obesity: Dietary interventions for weight management.
- Hypothalamic Dysfunction: Fluid goals, antidiuretics, growth hormone replacement, and thyroid medications.
- Hypoventilation: Tracheostomy and mechanical ventilation.
- Autonomic Dysregulation: Cardiac pacemaker, bowel medications, darkened room, and close monitoring of vital signs.

Anthropometrics

<table>
<thead>
<tr>
<th></th>
<th>10/12</th>
<th>Assessment</th>
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<tbody>
<tr>
<td>Weight</td>
<td>69.7kg (153.3lb)</td>
<td>89th%ile</td>
</tr>
<tr>
<td>Height</td>
<td>157.5cm (62in)</td>
<td>24th%ile</td>
</tr>
<tr>
<td>BMI</td>
<td>28.1</td>
<td>93rd%ile (overweight)</td>
</tr>
<tr>
<td>IBW</td>
<td>53-60kg</td>
<td></td>
</tr>
<tr>
<td>%IBW</td>
<td>116-132%</td>
<td>Overweight</td>
</tr>
<tr>
<td>UBW</td>
<td>69-72kg</td>
<td></td>
</tr>
<tr>
<td>Weight Changes</td>
<td>Stable x 1 mos</td>
<td>Non-significant, beneficial weight loss</td>
</tr>
<tr>
<td></td>
<td>-5.4% x 3 mos</td>
<td></td>
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<tr>
<td></td>
<td>-7.1% x 6 mos</td>
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Nutrition Care Plan

- PES (2/20): Involuntary weight gain r/t insatiable appetite, ROHHAD AEB hx significant weight gain x 1, 3, and 6 months.
- Goals: Wt stability or gradual wt loss to previous UBW of 68-70kg. Prevention of skin breakdown, adequate hydration, and energy for daily activities and therapies.

Clinical Observations

- Appeared overweight but not obese
- Energetic demeanor
- Behavioral disturbances
- History of polyphagia and polydipsia

Growth Charts

BMI-for-Age
Dietary History

<table>
<thead>
<tr>
<th>Date</th>
<th>Diet Order</th>
<th>Fluid Order</th>
<th>Reason for Change</th>
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<tbody>
<tr>
<td>6/14</td>
<td>900 kcals</td>
<td>Admit, restrict d/t hyperphagia</td>
<td></td>
</tr>
<tr>
<td>10/4</td>
<td>Regular pediatric: Ex: 2500mL</td>
<td>Changed d/t wt loss</td>
<td></td>
</tr>
<tr>
<td>8/27</td>
<td>1200 kcals Ex: 4500mL</td>
<td>ROHHAD dx</td>
<td></td>
</tr>
<tr>
<td>9/12</td>
<td>1200 kcals Snacks TID Ex: 2750mL</td>
<td>Current diet order</td>
<td></td>
</tr>
</tbody>
</table>

- PO intake = 100%
- Estimated needs (calculated using IBW 57kg): 1150-1440 kcals/day (20-25kcals/kg), 57-70g protein (1-1.2g/kg), 2750ml/day (to maintain fluid balance)

Analysis of Dietary Interventions

- Dietary interventions have been successful.
  - Current weight 69.7kg, meeting goal of 68-70kg.
  - Diet + multivitamin adequate to meet nutritional needs.
- Update PES statement: Overweight (NC-3.3.1) r/t ROHHAD AEB BMI 28.1 at 93rdiles BMI-for-age, hx of significant wt gain.
  - Continue current POC.
- Monitor weight, nutrition-related labs, skin status, PO intake, tolerance to diet order, and fluid balance and make recommendations PRN.

Prognosis & Conclusion

- High risk for sudden mortality.
- No treatment, but complications can be managed through symptom-targeted interventions.
- The patient is likely one of the oldest known living patients with ROHHAD.

References