BCCH GUIDELINES FOR ENDOCRINE NURSING CARE

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REGIONAL CLINIC REQUIREMENTS FOR ENDOCRINOLOGY

Facility: Available space for measuring children should provide comfort, privacy and opportunity for appropriate hand washing and other infection control measures between patients.

Equipment: Guidance for selection of appropriate measuring equipment is available through LHRSA/MCHB Training Module 4: Accurately Weighing and Measuring: Equipment
- Adult stadiometer and calibration rod
- Infant stadiometer and calibration rod
- Adult scales, broad based
- Infant scales
- Stool (flat top, stable base) for sitting height
- Blood pressure equipment including cuffs for all sizes
- Measuring tapes (plasticized)
- Calculator (metric converter preferred)

Paper Supplies:
- WHO Growth Charts for Canada (2014 revision, Set 2) appropriate to clinic patients: include body-mass index, head circumference, and syndrome-specific charts. May be supplied by hospital or downloaded.
- Ambulatory Nursing Notes
- Appropriate business cards

Endocrine Teaching Materials:
- Supply of frequently used forms
- Booklets, endocrine specific
- Online info, downloaded for specific patients
- Demonstration equipment: syringes; injection devices; blood glucose meters

Resource Materials for Nurses:
- Utilize BCCH Guidelines for Endocrine Clinic Nursing.
- Utilize Partners in Education “C” Binder and CD-ROM from the Pediatric Endocrinology Nursing Society.
- Utilize Clinical Handbook of Pediatric Endocrinology, 2nd ed. from CRC Press.
NURSING PREPARATION FOR ENDOCRINE CLINICS

Prior to clinic:
- Review charts.
- Ensure all forms are appropriate (growth charts, flow sheet).
- Ensure all documents are current and available (labs; bone age, etc).
- Plan for needed supplies, patient education materials, teaching time, forms.
- Arrange interpreter service if available.

Day of clinic:
- Calibrate equipment.

During the clinic:
- Check in patients (Receptionist); verify data.
- Lead the patient and family to the measuring area, introducing self, describing multidisciplinary team and the expected sequence of the visit.
- Ask the patient to change into light clothing for measurements.
- Plot growth, head circumference for children ≤5 yr; calculate growth velocity, BMI. Plot all appropriately.
- Apply EMLA® or Ametop® cream as needed.
- Assess current management of their endocrine condition, including medication schedule and administration methods.
- Determine learning needs. Document as needed.
- Communicate pertinent info to physician.
- Communicate with other services as needed (Social Worker; Community Health Nurse; Dietitian).

Follow Up:
- Administer medications as ordered. Ensure appropriate identification verification.
- Give medication instruction.
- Give stimulation test instructions.
- Update forms as needed (Hydrocortisone replacement forms annually and/or at each dosage change; copy to Health Records, Endocrinologist; Pediatrician, Family Doctor).
- Book additional nursing time (learning new skill; teleconferencing).
- Document as needed.
Between Appointments:
- Administer medications as ordered.
- Teach skills as needed.
- Contact family for further discussion and problem-solving.
- Document and notify Endocrinologist.

Documentation:
- Medications; dilution instructions; teaching; counselling; health management instructions should all be documented in the Health Record, and copied to the Endocrinologist.
- Telephone calls or other forms of communication such as e-mails should be documented in like manner.
## GROWTH

### SHORT STATURE

**Description:** A child with a height below the 5th centile for chronologic age (CA); growth velocity below the 3rd centile for CA; or height below the expected centile for mid-parental height. Possible etiology: familial short stature; constitutional delay of growth and puberty; chronic disease; association with a syndrome; poor nutrition; growth hormone deficiency.

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<th>Anthropometrics</th>
<th>Nursing Considerations</th>
<th>Documentation</th>
<th>Teaching</th>
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<tbody>
<tr>
<td>&quot;Standard Measurements&quot;</td>
<td>— Determine any medications which may affect growth</td>
<td>— Measurements: growth velocity; BMI (kg·m²)</td>
<td>— Description of growth assessment.</td>
</tr>
<tr>
<td>— Measure birth parents, first visit.</td>
<td>— Medications.</td>
<td>— Preparation for further testing</td>
<td>— Consider need for SW referral</td>
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</table>

**New Patient Information:**
1. Teaching materials about growth are available from the pharmaceutical companies who make growth hormone and also from growth support groups, listed on the EDU website.
GROWTH

GROWTH HORMONE DEFICIENCY

Description: Decreased or absent growth hormone production. Possible etiology: idiopathic; congenital abnormality of pituitary gland; damage to pituitary gland by tumour, trauma, infection, surgery, radiation or other treatments.

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<tr>
<td>— Standard Measurements.</td>
<td>— Determine any medications which may affect growth.</td>
<td>— Measurements; growth velocity; BMI (kg/m²); bone age</td>
<td>— Review skills for subcutaneous injections.</td>
</tr>
<tr>
<td>— Segment measurements and sitting height may be needed.(include link)</td>
<td>— Common hormone replacement: growth hormone</td>
<td>— Medications</td>
<td>— Review injection device as needed.</td>
</tr>
<tr>
<td>— Measure birth parents, first visit.</td>
<td></td>
<td>— Growth hormone: Note management of injections; dilution instructions.</td>
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</table>

New Patient Information:
2. Provide family with BCCH handout Starting Growth Hormone.
3. Advise family to confirm Fair PharmaCare registration (BC); utilize pharmaceutical company’s Reimbursement Assistance Program, if needed, for assistance with the deductible portion.
4. Utilize Teaching Checklist: Growth Hormone: (Pens) or Teaching Checklist: Growth Hormone: (Vials).
5. Give suitable info from BCCH website.
GROWTH

TALL STATURE

Description: A child with a height above the 97th centile for chronologic age (CA), that is, 2 standard deviations (SD) above the mean. Possible etiology: familial tall stature; early puberty; obesity; association with a syndrome; growth hormone excess.

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<tr>
<td>— Standard Measurements</td>
<td>— Determine any medications which may affect growth</td>
<td>— Measurements: growth velocity; BMI (kg/m²)</td>
<td>— Description of growth assessment.</td>
</tr>
<tr>
<td>— Measure birth parents, first visit.</td>
<td>— Medications.</td>
<td>— Consider need for SW referral</td>
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New Patient Information:
The Tall Book by Arianne Cohen (sample available in clinic, parents may order from Chapters or Amazon).
THYROID

HYPOTHYROIDISM

Description: Deficiency of thyroid hormones. Possible etiology: Congenital; acquired: autoimmune disorder (Hashimoto's thyroiditis); post ablation therapy for hyperthyroidism; secondary to hypopituitarism

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<th>Teaching</th>
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<tr>
<td>— Standard Measurements</td>
<td>— Common hormone replacement: levothyroxine</td>
<td>— Measurements: growth velocity; BMI (kg/m²)</td>
<td>— Review replacement therapy and age related issues:</td>
</tr>
<tr>
<td>— Segments not needed</td>
<td>— Medications.</td>
<td></td>
<td>- medication schedule</td>
</tr>
<tr>
<td>— Measure birth parents, first visit.</td>
<td></td>
<td>— method of administration</td>
<td>— Review relevance of labs</td>
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New Patient Information:
1. Give BCCH booklet Hypothyroidism.
2. Give suitable info from BCCH website.
3. Demonstrate pill cutter and crusher.
4. Advise clear 7-day pill holder.
5. Discuss potential problems: pill admin to infant; spitting up/ replacing missed pills; timing of lab work.
6. Support groups: Thyroid Foundation of Canada; American Thyroid Association.
THYROID

HYPERTHYROIDISM

Description: Excess production of thyroid hormones by the thyroid gland.

Possible Etiology: Graves disease; subacute thyroiditis; hyperthyroid phase of Hashimoto thyroiditis; tumor.

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<tr>
<td>— Standard Measurements — Segments not needed — Measure birth parents, first visit.</td>
<td>— Review medications - propylthiouracil (PTU®) - methimazole (Tapazole®) - propranolol (short-term)</td>
<td>— Measurements: growth velocity; BMI (kg/m²) — Adherence to prescribed therapy</td>
<td>— Review anti-thyroid drug therapy, importance of adherence and regular labs — Review reporting of side effects promptly — Review illness management</td>
</tr>
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New Patient Information:
1. Give BCCH booklet Hyperthyroidism (also available in Chinese).
2. Give suitable info from BCCH website.
3. Clarify exercise limitations.
4. Advise clear plastic 7-day pill holder.
5. Confirm instructions for illness management and possible side effects of anti-thyroid drug therapy.
6. Support groups: Thyroid Foundation of Canada; National Graves Disease Foundation; American Thyroid Association.
ADRENAL

ADRENAL INSUFFICIENCY

Description: Insufficient amount of the adrenal hormone, cortisol, to enable cellular function. Cortisol is one of the hormones that sustains blood pressure and blood sugar in the normal range.

Possible Etiology: Hypopituitarism; congenital adrenal hyperplasia; Addison disease; post-adrenalectomy; adrenal suppression from high-dose glucocorticoid use.

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<tr>
<td>— As per specific condition</td>
<td>— Common hormone replacement: one of the following glucocorticoids: - hydrocortisone (Cortef®) - prednisolone (Pediapred®) - prednisone</td>
<td>— As per specific condition. — Utilize Learning Pathway: Adrenal Insufficiency for ongoing recording. — Illness management details. — Medical alert? — Flu shot?</td>
<td>— Age-specific anticipatory guidance for illness management. — Review IM skills annually. — Repeat teaching for alternate caregivers.</td>
</tr>
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</table>

New Patient Information:
1. Inpatient: Ensure BCCH Reference Care Plan: Adrenal Insufficiency is available.
2. Give diagnosis-specific information (see guidelines for specific condition in following pages).
5. Utilize Management of Hydrocortisone Replacement annually and for dosage change.
6. Utilize Endocrine Travel Letter.
7. Utilize School Letter for the Cortisol-Dependent Student.
8. Utilize Community Health Services Referral Form for Children who are Cortisol-Dependent prior to school entry.
9. Utilize Influenza Vaccine for Children Who are Cortisol-Dependent.
10. Utilize Immunizations for Children who are Cortisol-Dependent.
11. Medical alert to say “Adrenal Insufficiency: steroid-dependent”.

— As per specific condition
ADRENAL

ADDISON DISEASE
See Guidelines for Adrenal Insufficiency

Description: Autoimmune disease, destroying the cells of the adrenal cortex. The adrenal gland is then not able to produce glucocorticoid or mineralocorticoid.

Possible Etiology: Unknown

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<td>— Standard Measurements</td>
<td>— Review medications:</td>
<td>— As per Guidelines for Adrenal Insufficiency</td>
<td>— As per Guidelines for Adrenal Insufficiency</td>
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<tr>
<td>— Measure birth parents, first visit.</td>
<td>- Glucocorticoids</td>
<td></td>
<td>— Florinef® function.</td>
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<td></td>
<td>- Florinef®</td>
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New Patient Information: Diagnosis likely known prior to clinic appointment.

1. Determine what the family knows and what they are hoping for.
2. Utilize Teaching Checklist: Hydrocortisone Replacement and other materials described in Guidelines for Adrenal Insufficiency.
3. Give suitable info from BCCH website.
5. Support group: Canadian Addison Society; National Adrenal Diseases Foundation.
CONGENITAL ADRENAL HYPERPLASIA

See Guidelines for Adrenal Insufficiency

Description: Inherited disorder marked by deficiency or absence of one or two adrenal hormones (cortisol and aldosterone). May result in overproduction of adrenal androgens. May be a severe or mild deficiency.

Possible Etiology: Genetic condition, autosomal recessive, leading to a deficiency of enzymes essential for production of adrenal hormones.

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| — Standard Measurements | — Review medications:  
- Glucocorticoid
- Florinef® (most patients)
- Salt (most infants)  
— Accurate BP necessary for management of Florinef®, salt | — As per Guidelines for Adrenal Insufficiency  
— Management of salt | — As per Guidelines for Adrenal Insufficiency  
— Florinef® function |

New Patient Information:
1. Inpatient: Ensure BCCH Reference Care Plan: Adrenal Insufficiency is available to bedside nurse.
3. Give suitable info from BCCH website.
4. Utilize Teaching Checklist: Hydrocortisone Replacement and other materials described in Guidelines for Adrenal Insufficiency.
5. Ensure that family understands Salt Replacement for Mineralocorticoid Deficiency.
ADRENAL

ADRENAL SUPPRESSION
See Guidelines for Adrenal Insufficiency

Description: Inability of the adrenal glands to produce glucocorticoid as a result of suppression of the hypothalamic pituitary adrenal axis.

Possible Etiology: High-dose glucocorticoids may be given to treat an underlying condition. This will suppress the axis while they are being used, during weaning and for an unknown length of time after being discontinued. The individual will be vulnerable to adrenal crisis during illness episodes if the exogenous glucocorticoid is less than the body requires.

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<td>— Standard Measurements</td>
<td>— Review medications:</td>
<td>As per Guidelines for Adrenal Insufficiency</td>
<td>As per Guidelines for Adrenal Insufficiency</td>
</tr>
<tr>
<td>— Measure birth parents, first visit.</td>
<td>— Glucocorticoid</td>
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New Patient Information: Diagnosis likely known prior to clinic appointment.
1. Determine the relevance of Guidelines for Adrenal Insufficiency and materials for this particular situation. IM skills, medical alert, and some other teaching may not be necessary.
2. Utilize Teaching Checklist: Hydrocortisone Replacement and other materials described in Guidelines for Adrenal Insufficiency, as needed.
3. Utilize Illness Management During Adrenal Suppression handout.
4. Give suitable info from BCCH website.
HYPOPITUITARISM
(See Guidelines for Adrenal Insufficiency)

Description: Decrease or absence of one or more pituitary hormones. Panhypopituitarism is the absence of all pituitary hormones.

Possible Etiology: Congenital abnormality; acquired as a result of tumor; and related treatments such as surgery, radiation; trauma; infection.

New Patient Information:
1. Inpatient: Ensure BCCH Reference Care Plan: Adrenal Insufficiency and BCCH Reference Care Plan: Diabetes Insipidus are available.
2. Give BCCH booklet Hypopituitarism; also Diabetes Insipidus booklet for affected infants.
3. Give suitable info from BCCH website.
5. Utilize Teaching Checklist: Growth Hormone: (Pens) or Teaching Checklist: Growth Hormone: (Vials).
6. Support groups: Panhypopituitarism from the MAGIC Foundation; Pituitary Tumor Network Association.
PITUITARY

DIABETES INSIPIDUS (CENTRAL)

Description: Excess fluid loss via the kidneys as a result of inadequate amount of antidiuretic hormone (ADH) normally produced in the hypothalamus and posterior pituitary gland. This is not to be confused with nephrogenic diabetes insipidus, a renal condition.

Possible Etiology: Congenital abnormality; acquired as a result of damage to the posterior pituitary gland (head injury, surgery, tumor, infection, radiation).

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<td>— Standard Measurements</td>
<td>— Review medications</td>
<td>— Measurements; growth velocity; BMI (kg/m²)</td>
<td>— Review administration of intranasal med as needed.</td>
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<tr>
<td>— Measure birth parents first visit</td>
<td>— DDAVP® nasal formulation; tablet</td>
<td>— Medications</td>
<td>— Problem-solve “breakthrough” issues at school.</td>
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<td>— Discuss “breakthrough”</td>
<td>— Wearing medical alert</td>
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<td>— Provide Fluid Balance Record as needed</td>
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<td>— Provide Diabetes Insipidus Letter as needed</td>
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New Patient Information:
1. Inpatient: Ensure BCCH Reference Care Plan: Diabetes Insipidus is available to bedside nurses.
2. Give BCCH booklet Diabetes Insipidus.
3. Give suitable info from BCCH website.
4. Teach relevant skills: measuring ins and outs; daily weights; test strips for specific gravity; administration of med via rhinyle.
5. Review management of illness episodes.
### DELAYED PUBERTY

**Description:** Lack of secondary sex characteristics by age 14 for a boy, or age 13 for a girl.

**Possible Etiology:** Constitutional delay—late bloomer; systemic illness such as heart disease; eating disorders; inadequate function of hypothalamus, pituitary or gonads. Hypothalamic, pituitary or gonadal dysfunction may indicate permanent hypogonadism.

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<th><strong>Documentation</strong></th>
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</tr>
</thead>
<tbody>
<tr>
<td>— <em>Standard Measurements</em> — Measure birth parents first visit.</td>
<td>— Review medications which may impact growth and development.</td>
<td>— Measurements; growth velocity; BMI (kg/m²); bone age — Medications</td>
<td>— Booklet for constitutional delay in boys: <em>This is Slow Growing</em> (out of print 2010). — Handout: <em>Testosterone Injections</em>, as needed for boys.</td>
</tr>
</tbody>
</table>

**New Patient Information:**
1. Give suitable info from BCCH website.
2. See *Guidelines for Hypogonadism (Sex-Hormone Deficiency)* as needed.
PUBERTY

HYPOGONADISM (SEX-HORMONE DEFICIENCY)

Description: Permanent lack of age-appropriate sex hormones and therefore lack of secondary sex characteristics and fertility challenges.

Possible Etiology: Gonadal dysfunction or lack of hypothalamic or pituitary stimulating hormones; usually permanent; occasionally temporary, caused by malnutrition or chronic disease.

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<tbody>
<tr>
<td>— Standard Measurements</td>
<td>— Hormone replacement, age- and sex-specific. — Testosterone injections or topical gel — Estrogen and progesterone</td>
<td>— Measurements; growth velocity; BMI (kg/m²); bone age — Medications</td>
<td>— BCCH booklet for hormone replacement therapy: It’s All Chemistry For Girls/For Boys. — Handout: Testosterone Injections, as needed for boys.</td>
</tr>
</tbody>
</table>

New Patient Information:
1. Determine what the family knows and collaborate with them in disclosure with their child. Document and follow this aspect of communication.
2. Give diagnosis specific information (e.g. Turner syndrome, Kallman syndrome).
3. Give suitable info from BCCH website.
4. Give booklet pertaining to appropriate hormone replacement therapy.
5. Teach intramuscular administration (testosterone) as needed.
6. Ensure reproductive information is made available to family. Assisted reproductive technology may be appropriate in some circumstances.
7. Ensure counselling opportunity as needed.
PUBERTY

CENTRAL PREOCIOUS PUBERTY

Description: Development of secondary sex characteristics before the age of 9 for a boy or 7 for a girl.

Possible Etiology: Precocious puberty may be central (premature maturation of the hypothalamic-pituitary-gonadal axis); constitutional; caused by CNS tumours or treatments; idiopathic. Only central precocious puberty is treated by a GnRH analog.

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<tr>
<td>— Standard Measurements</td>
<td>— Common therapy: GnRH analog: Lupron Depot® or Decapeptyl®.</td>
<td>— Measurements; growth velocity; BMI (kg/m²); bone age</td>
<td>— Preparation for stimulation testing</td>
</tr>
<tr>
<td>— Measure birth parents first visit.</td>
<td>— Review medication schedule.</td>
<td>— Medications</td>
<td>— Teach parent IM administration if desired.</td>
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<td></td>
<td></td>
<td>— Provide Injection Schedule</td>
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New Patient Information:
1. Give appropriate information: Precocious Puberty from the MAGIC Foundation; toosoon.com; Abbott information booklet; Starting Lupron Depot®.
2. Give suitable info from BCCH website.
4. Discuss side effects, expectations.
5. Provide Injection Schedule and assist with scheduling.
6. Provide opportunity for discussion of psychosocial concerns.
CALCIUM DISORDERS

HYPOPARATHYROIDISM

Description: A deficiency of parathyroid hormone secretion by the parathyroid gland, causing hypocalcemia, and hyperphosphatemia.

Possible Etiology: Genetic disorder, 22q deletion; surgical excision; autoimmune disorder; complication of thalassemia treatment.

Pseudohypparathyroidism: Similar manifestation and treatment, but caused by cellular resistance to PTH.

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<tr>
<td>— Standard Measurements — Measure birth parents first visit.</td>
<td>— Review medications — Calcium — Calcitriol (Rocaltrol®)</td>
<td>— Measurements; growth velocity; BMI (kg/m²); bone age — Medications</td>
<td>— Review medication schedule and method of administration. Discuss calcium replacements.</td>
</tr>
</tbody>
</table>

New Patient Information:
1. Give suitable info from BCCH website.
2. Review implications of low calcium.
HYPOGLYCEMIC CONDITIONS

KETOTIC HYPOGLYCEMIA

**Description:** Low blood sugar (below 3.0 mmol/L), unexplained by other causes (GHD; low cortisol); insulin levels are not high; ketones are present in the blood and urine.

**Possible Etiology:** Prolonged fast; dietary interruption

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<th>Anthropometrics</th>
<th>Nursing Considerations</th>
<th>Documentation</th>
<th>Teaching</th>
</tr>
</thead>
</table>
| — *Standard Measurements*  
— Measure birth parents first visit. | — Review medications | — Measurements; growth velocity; BMI (kg/m²); bone age  
— Medications | — BG testing |

**New Patient Information:**
1. Ensure Dietitian consult.
2. Teach BG testing using monitor.
5. Give suitable info from BCCH website.
HYPOGLYCEMIC CONDITIONS

HYPERINSULINISM

Description: Rare condition of frequent hypoglycemia accompanied by high insulin levels.

Possible Etiology: Gene mutation of chromosome 11, which results in dysregulation of insulin secretion

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<td>— Review medications:</td>
<td>— Measurements; growth velocity; BMI (kg/m²); bone age</td>
<td>— BG testing</td>
</tr>
<tr>
<td>— Measure birth parents first visit.</td>
<td>- Diazoxide®</td>
<td>— Medications</td>
<td>— Review subcutaneous technique</td>
</tr>
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<td></td>
<td>- Sandostatin®</td>
<td></td>
<td>— Create individualized</td>
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<tr>
<td></td>
<td>- Polycose® (infants)</td>
<td></td>
<td>— Individualized hypoglycemia management instructions</td>
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<td></td>
<td>- Cornstarch (toddlers)</td>
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</tbody>
</table>

New Patient Information:
1. Ensure social work consult for At Home Program application as needed.
2. Ensure Dietitian consult.
3. Teach BG testing.
5. Teach subcutaneous injection technique as needed.
6. Teach Glucagon for Severe Hypoglycemia in Hyperinsulinism and/or Mini-Dose Glucagon for Preventing Severe Hypoglycemia as needed.
7. Ensure NG teaching has been done as needed.
8. Give suitable info from BCCH website.
SYNDROMES

KLINFELETER SYNDROME (47,XXY MALE)

Description: Genetic condition as a result of an extra X chromosome (rarely more than one extra X.) Hypogonadism usually leads to insufficient testosterone production, fertility problems. May include some difficulties with language skills and social skills.

Possible Etiology: Unknown

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<tbody>
<tr>
<td>— <em>Standard Measurements</em></td>
<td>— Review medications: testosterone replacement as needed.</td>
<td>— Measurements, growth velocity, BMI (kg/m²), bone age</td>
<td>— Booklet: <em>It's All Chemistry For Boys</em> prior to puberty.</td>
</tr>
<tr>
<td>— Measure birth parents first visit.</td>
<td></td>
<td>— Medications</td>
<td>— Handout: <em>Testosterone Injections</em> as needed</td>
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<tr>
<td></td>
<td></td>
<td>— Disclosure status</td>
<td>— Teach IM technique PRN</td>
</tr>
</tbody>
</table>

New Patient Information: Diagnosis likely known prior to clinic appointment.

1. Determine what the family knows and what they are hoping for.
2. Give suitable info from BCCH website.
3. Determine parents' plan for disclosure, and support their plan (document clearly—follow up by phone as needed).
### SYNDROMES

#### PRADER–WILLI SYNDROME

**Description:** Genetic condition with the following clinical findings: hypotonia and feeding difficulties in infancy; hyperphagia; hypogonadism; short stature; dysmorphic facial features; developmental disabilities.

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<tbody>
<tr>
<td>— Standard Measurements</td>
<td>— Medications:</td>
<td>— Measurements, growth velocity, BMI (kg/m²), bone age</td>
<td>— Community Health Services referral prior to school.</td>
</tr>
<tr>
<td>— Include PWS growth chart</td>
<td>— growth hormone (not a BC PharmaCare benefit)</td>
<td>— Medications</td>
<td>— HRT as needed</td>
</tr>
<tr>
<td>— Measure birth parents first visit.</td>
<td>— sex-hormone replacements</td>
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</tbody>
</table>

**New Patient Information:** Diagnosis likely known prior to clinic appointment.
1. Determine what the family knows and what they are hoping for.
2. Give suitable info from BCCH website.
3. Suggest **BCCH Family Support and Resource Centre DVD:** *Food, Behavior and Beyond: Practical Management for the Child and Adult with PWS* (also available through Prader–Willi Syndrome Association USA).
4. Utilize **Teaching Checklist: Growth Hormone: (Pens)** or **Teaching Checklist: Growth Hormone: (Vials)**, as needed.
5. Support groups: **British Columbia Prader Willi Syndrome Association; Foundation for Prader–Willi Research Canada; the Prader–Willi Syndrome Association USA** offers a free info package.
## SYNDROMES

### RUSSELL-SILVER SYNDROME

**Description:** Genetic condition with the following clinical findings: intrauterine growth retardation (IUGR); hypoglycemia in infancy; short stature; feeding difficulties; dysmorphic facial features.

**Possible Etiology:** genetic

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<tbody>
<tr>
<td>— Standard Measurements</td>
<td>— Hypoglycemia management</td>
<td>— Measurements: growth velocity, BMI (kg/m²), bone age</td>
<td>— Blood glucose monitoring</td>
</tr>
<tr>
<td>— Include Russell-Silver growth chart</td>
<td>— Growth hormone (not a BC PharmaCare benefit)</td>
<td>— Medications</td>
<td></td>
</tr>
<tr>
<td>— Measure birth parents first visit.</td>
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</table>

**New Patient Information:** Diagnosis likely known prior to clinic appointment.

1. Determine what the family knows and what they are hoping for.
2. Give suitable info from BCCH website.
3. Show the following if interested: *Russell-Silver Syndrome (MAGIC Foundation).*
4. Utilize *Teaching Checklist: Growth Hormone: (Pens)* or *Teaching Checklist: Growth Hormone: (Vials)*, as needed.
5. Support groups: *Russell-Silver Support; MAGIC Foundation.*
**SYNDROMES**

**TURNER SYNDROME**

**Description:** Genetic condition caused by the absence of all or part of the second X chromosome in some or all of the cells of the body. Typically the affected girl has short stature and hypogonadism which results in insufficient sex hormone production and fertility problems. May include some difficulties with language skills and social skills.

**Possible Etiology:** Unknown

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<td>— <em>Standard Measurements</em> —</td>
<td>— Review medications: —</td>
<td>— Measurements, growth velocity, BMI (kg/m²), bone age</td>
<td>— Booklet: It's All Chemistry For Girls, prior to puberty.</td>
</tr>
<tr>
<td>— Include TS growth chart</td>
<td>— sex-hormone replacement as needed.</td>
<td>— Medications</td>
<td></td>
</tr>
<tr>
<td>— Measure birth parents first visit.</td>
<td>— growth hormone (not a BC PharmaCare benefit)</td>
<td>— Disclosure status</td>
<td></td>
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</table>

**New Patient Information:** Diagnosis likely known prior to clinic appointment.

1. Determine what the family knows and what they are hoping for.
2. Give suitable info from BCCH website.
4. Utilize *Teaching Checklist: Growth Hormone: (Pens)* or *Teaching Checklist: Growth Hormone: (Vials)*, as needed.
5. Determine parents’ plan for disclosure, and support their plan (document clearly— follow up by phone as needed).
6. Support groups: *BC Turner Syndrome Network; Turner Syndrome Society of Canada; Turner Syndrome Society of the US.*