ENDOCRINOLOGY & DIABETES UNIT
Endocrinology Clinic: 604-875-3611
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http://endodiab.bcchildrens.ca

BCCH GUIDELINES FOR ENDOCRINE NURSING CARE

www.bcchildrens.ca/endocrinology-diabetes-site/documents/endorncare.pdf

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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*.
- Regional Clinic forms, BCCH forms, available at http://endodiab.bcchildrens.ca.
- 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.

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents, first visit.	— Determine any medications which may affect growth	— Measurements: growth velocity; BMI ($kg \div m^2$) — Medications.	— Description of growth assessment. — Preparation for further testing — Consider need for SW referral

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.
2. Booklet: *Patterns of Growth* from the Human Growth Foundation.
3. Support groups: Human Growth Foundation; MAGIC Foundation.

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.

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

New Patient Information:

1. Prepare family for child to start GH, using product-specific 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.

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents, first visit.	— Determine any medications which may affect growth	— Measurements: growth velocity; BMI (kg/m^2) — Medications.	— Description of growth assessment. — Consider need for SW referral

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

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Segments not needed — Measure birth parents, first visit.	— Common hormone replacement: levothyroxine	— Measurements: growth velocity; BMI (kg/m^2) — Medications.	— Review replacement therapy and age related issues: - medication schedule - method of administration — Review relevance of labs

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.

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Segments not needed — Measure birth parents, first visit.	— Review medications - propylthiouracil (PTU®) - methimazole (Tapazole®) - propranolol (short-term)	— Measurements: growth velocity; BMI (kg/m^2) — Adherence to prescribed therapy	— Review anti-thyroid drug therapy, importance of adherence and regular labs — Review reporting of side effects promptly — Review illness management

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.

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

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).
3. Utilize *Learning Pathway: Adrenal Insufficiency*.
4. Utilize *Teaching Checklist: Hydrocortisone Replacement*.
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"**.

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

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents, first visit.	— Review medications: - Glucocorticoids - Florinef®	— As per *Guidelines for Adrenal Insufficiency*	— As per *Guidelines for Adrenal Insufficiency* — Florinef® function.

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.
4. Best info (lengthy): *Addison's Disease Owner's Manual*.
5. Support group: Canadian Addison Society; National Adrenal Diseases Foundation.

ADRENAL

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.

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents, first visit.	— 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.
2. Give BCCH Booklet *Congenital Adrenal Hyperplasia*; show book *Congenital Adrenal Hyperplasia: A Parent's Guide* (by Hsu CY & Rivkees SA) available through a general bookstore.
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*.
6. Additional information for families of virilized females: *How the Body Works: Genital Development* (Hospital for Sick Children); *Consortium on the Management of Disorders of Sex Development: Handbook for Parents*.
7. Support group: Congenital Adrenal Hyperplasia Research Education & Support (CARES).

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.

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents, first visit.	— Review medications: — Glucocorticoid	As per *Guidelines for Adrenal Insufficiency*	— As per *Guidelines for Adrenal Insufficiency*

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.

PITUITARY

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.

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents first visit	— Review hormone replacements: - glucocorticoid - levothyroxine - growth hormone - sex steroids during adolescence - DDAVP®	— Measurements: growth velocity; BMI (kg/m^2) — As per *Guidelines for Adrenal Insufficiency* — Management of growth hormone — Management of DDAVP® — wearing medical alert	— As per *Guidelines for Adrenal Insufficiency* — As per *Guidelines For Growth Hormone Deficiency* — As per *Guidelines for Central Diabetes Insipidus* — As per *Guidelines for Hypothyroidism*

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.
4. Utilize *Teaching Checklist: Hydrocortisone Replacement*.
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).

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents first visit	— Review medications — DDAVP® nasal formulation; tablet — Discuss "breakthrough" — Provide *Fluid Balance Record* as needed — Provide *Diabetes Insipidus Letter* as needed.	— Measurements; growth velocity; BMI (kg/m^2) — Medications — Wearing medical alert	— Review administration of intranasal med as needed. — Problem-solve "breakthrough" issues at school.

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.
6. Support Group: Diabetes Insipidus Foundation.

PUBERTY

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.

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents first visit.	— Review medications which may impact growth and development.	— Measurements; growth velocity; BMI (kg/m^2); bone age — Medications	— Booklet for constitutional delay in boys: *This is Slow Growing* (out of print 2010). — Handout: *Testosterone Injections*, as needed for boys.

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.

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents first visit.	— Hormone replacement, age- and sex-specific. — Testosterone injections or topical gel — Estrogen and progesterone	— Measurements; growth velocity; BMI (kg/m^2); bone age — Medications	— BCCH booklet for hormone replacement therapy: *It's All Chemistry For Girls/For Boys.* — Handout: *Testosterone Injections,* as needed for boys.

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 PRECOCIOUS 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.

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents first visit.	— Common therapy: GnRH analog: Lupron Depot® or Decapeptyl®. — Review medication schedule.	— Measurements; growth velocity; BMI (kg/m^2); bone age — Medications — Provide *Injection Schedule*	— Preparation for stimulation testing — Teach parent IM administration if desired.

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.
3. Discuss process for beginning treatment.
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.

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

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents first visit.	— Review medications — Calcium — Calcitriol (Rocaltrol®)	— Measurements; growth velocity; BMI (kg/m^2); bone age — Medications	— Review medication schedule and method of administration. Discuss calcium replacements.

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

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents first visit.	— Review medications	— Measurements; growth velocity; BMI (kg/m^2); bone age — Medications	— BG testing

New Patient Information:

1. Ensure Dietitian consult.
2. Teach BG testing using monitor.
3. Review *Hypoglycemia in the Endocrine Patient* instructions.
4. Clarify use of *Blood Glucose Log for Endocrine Patients*; faxing instructions.
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

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents first visit.	— Review medications: - Diazoxide® - Sandostatin® - Polycose® (infants) - Cornstarch (toddlers)	— Measurements; growth velocity; BMI (kg/m^2); bone age — Medications	— BG testing — Review subcutaneous technique — Create individualized — Individualized hypoglycemia management instructions

New Patient Information:

1. Ensure social work consult for At Home Program application as needed.
2. Ensure Dietitian consult.
3. Teach BG testing.
4. Clarify use of *Blood Glucose Log for Hyperinsulinism*; faxing instructions.
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.
9. Support group: sur1.org; Congenital Hyperinsulinism International.

SYNDROMES

KLINEFELTER 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

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Measure birth parents first visit.	— Review medications: testosterone replacement as needed.	— Measurements, growth velocity, BMI (kg/m^2), bone age — Medications — Disclosure status	— Booklet: *It's All Chemistry For Boys* prior to puberty. — Handout: *Testosterone Injections* as needed — Teach IM technique PRN

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).
4. Support groups: American Association for Klinefelter Syndrome Information and Support; KS&A: Knowledge, Support & Action; Klinefelter's Syndrome Association UK.

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.

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Include PWS growth chart — Measure birth parents first visit.	— Medications: - growth hormone (not a BC PharmaCare benefit) - sex-hormone replacements	— Measurements, growth velocity, BMI (kg/m^2), bone age — Medications	— Community Health Services referral prior to school. — HRT as needed

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

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Include Russell-Silver growth chart — Measure birth parents first visit.	— Hypoglycemia management — Growth hormone (not a BC PharmaCare benefit)	— Measurements: growth velocity, BMI (kg/m^2), bone age — Medications	— Blood glucose monitoring

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

Anthropometrics	Nursing Considerations	Documentation	Teaching
— *Standard Measurements* — Include TS growth chart — Measure birth parents first visit.	— Review medications: - sex-hormone replacement as needed. - growth hormone (not a BC PharmaCare benefit)	— Measurements, growth velocity, BMI (kg/m^2), bone age — Medications — Disclosure status	— Booklet: *It's All Chemistry For Girls*, prior to puberty.

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. Provide the following if interested: *Turner Syndrome: Across the Lifespan*; *Turner Syndrome Transition Passport*; *Turner Syndrome: A Guide for Families*.
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.