orphanaesthesia

Anaesthesia recommendations for patients suffering from

Congenital hypothyroidism

Disease name: Congenital Hypothyroidism

ICD 10: E03.0 with diffuse goiter

E03.1 without goiter

Synonyms: CH, neonatal hypothyroidism, fetal iodine deficiency disorder

Congenital hypothyroidism is a common preventable cause of mental retardation. The overall incidence is approximately 1:4,000. Females are affected about twice as often as males. Approximately 85% of cases are sporadic, while 15% are hereditary. The most common sporadic etiology is thyroid dysgenesis, with ectopic glands being more common than aplasia or hypoplasia. In untreated patients, symptoms and signs include the decreased activity, large anterior fontanelle, poor feeding, short stature or failure to thrive, jaundice, decreased stooling or constipation, hypotonia, and hoarse cry. The physical findings of hypothyroidism may or may not be present at birth. Signs include the coarse facial features, macroglossia, large fontanelles, umbilical hernia, mottled, cool, and dry skin, developmental delay, pallor, myxedema, goiter. In patients properly treated, there are no clinical signs.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

---

Disease summary

While the pathogenesis of dysgenesis is largely unknown, some cases are now discovered to be the result of mutations in the transcription factors PAX-8, FOXE1 (TTF-2), NKX2-1 (TTF-1), NKX2-5, GLIS3, and others. Loss of function mutations in the thyrotropin (TSH) receptor have been demonstrated to cause some familial forms of athyreosis. The most common hereditary etiology is the inborn errors of thyroxine (T4) synthesis. Recent mutations have been described in the genes coding for the sodium/iodide symporter, thyroid peroxidase (TPO), and thyroglobulin. Transplacental passage of a maternal thyrotropin receptor blocking antibody (TRB-Ab) causes a transient form of familial congenital hypothyroidism. The vast majority of infants are now diagnosed after detection through newborn screening programs using a primary T4-backup TSH or primary TSH test. Screening test results must be confirmed by serum thyroid function tests. Thyroid scintigraphy, using 99mTc or 123I, is the most accurate diagnostic test to detect thyroid dysgenesis or one of the inborn errors of T4 synthesis. Thyroid sonography is nearly as accurate, but it may miss some cases of ectopic glands. If maternal antibody-mediated hypothyroidism is suspected, measurement of maternal and/or neonatal TRB-Ab will confirm the diagnosis. The goals of treatment are to raise the serum T4 as rapidly as possible into the normal range, adjust the levothyroxine dose with growth to keep the serum T4 (or free T4) in the upper half of the normal range and the TSH normal, and maintain normal growth and development while avoiding overtreatment. An initial starting dose of 10-15 µg/kg per day is recommended; this dose will decrease on a weight basis over time. Serum T4 (or free T4) and TSH should be monitored every 1-2 months in the first year of life and every 2-3 months in the second and third years, and less frequently thereafter.

Rarely, congenital hypothyroidism is due to pituitary deficiency. In this case, other pituitary hormones, overall GH and ACTH, may be undetectable causing hypoglycaemia and adrenal insufficiency.

Typical surgery

In infants with congenital hypothyroidism, extra thyroidal congenital malformations have a prevalence of 8.4%. Of these, the majority are cardiac (e.g. atrial septal defect with atrioventricular conduction defect in patients with *NKX2-5* mutations). Cleft lip and cleft palate may be present in congenital hypothyroid neonates, as well as other midline defects.

A mutation in *FOXE1* causes a syndrome of thyroid dysgenesis, choanal atresia, cleft palate, bifid epiglottis, and spiky hair also known as Bamforth-Lazarus syndrome. *NKX2-1* (*TTF-1*) mutations may be associated with respiratory distress and choreoathetosis (brain-lung-thyroid syndrome), and *GLIS3* mutations may be associated with congenital diabetes mellitus and glaucoma.

Thyroidectomy is not recommended for congenital hypothyroidism. However, patients with congenital hypothyroidism may require surgery for other reasons e.g diffuse goiter.

Type of anaesthesia

Regional anaesthesia appears to be safe because it avoids all the airway related complications, and it is the choice of anaesthesia if the level of surgery permits its use, however one should anticipate precipitous hypotension and postop shivering

General anaesthesia; there are reports of thyroid surgeries done with cervical plexus block and with LMA with spontaneous ventilation endotracheal intubation with a flexometallic tube will be the safest option.

Necessary additional diagnostic procedures (preoperative)

Patients with subclinical hypothyroidism pose no problems during anaesthesia and it is not necessary to initiate thyroxin preoperatively in these groups. In the mild-moderate hypothyroidism there are no controlled studies to favour preoperative thyroxin therapy except a few case reports, hence preoperatively thyroxin therapy in these groups is tailored to the prevailing circumstances. In severe hypothyroidism (T4≤ 1µg/dl) for elective surgeries, surgery is deferred until euthyroid state is achieved. Emergency surgery in this group is risky because of anticipated cardiovascular instability and myxoedema coma. In these cases, intravenous thyroxine using age-appropriate dosing (in consultation with a pediatric endocrinologist) with ECG monitoring should be considered.

Thyroxine supplement has to be continued until the morning of surgery. Antisialogogue and antiemetic premedication is helpful since these patients have decreased gastrointestinal tract motility and bradycardia. It is better to avoid sedative and narcotic premedication.

Particular preparation for airway management

While assessing the patient, apart from the routine evaluation, attention has to be paid to the airway management.

In the literature, one study claimed that the rate of difficult airway is 11.1%

In patients with CH, especially in untreated subjects, the anaesthesiologist should be prepared for a difficult airway and anaesthetic management requiring close observation both preoperatively and postoperatively.

Other problems to be anticipated are airway problems due to macroglossia, upper airway edema, and airway deviation due to goiter.

Particular preparation for transfusion or administration of blood products

While assessing the patient, apart from routine evaluation, attention has to be paid to anaemia and heart defects.

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transport or mobilisation

Early mobilisation recommended for prophylaxis of thromboembolism.

Probable interaction between anaesthetic agents and patient's long-term medication

Hypothyroid patients are supposed to be more sensitive to anaesthetic drugs and inhalational agents though there is no clinical evidence supporting this.

Generally, these patients use thyroxine. There is no evidence for its interaction with anaesthetic agents.

Anaesthesiologic procedure

Hypothyroid patients are supposed to be more sensitive to anaesthetic drugs and inhalational agents though there is no clinical evidence supporting this.

The observed decrease in the MAC value for inhaled anesthetics is not clinically significant and probably due to decreased cardiac output and blood volume, as well as decreased metabolism and excretion.

Ketamine is the better induction agent in hypothyroidism because it will not produce hypotension and bradycardia.

Barbiturates and benzodiazepines can also be used if ketamine is contraindicated and hypothyroidism is mild or well controlled.

Anaesthesia is best maintained with oxygen and nitrous oxide and with intermittent opioids and muscle relaxants.

Inhalational agents are better avoided or used very cautiously.

Particular or additional monitoring

Pulse oximeter, NIBP, ECG, temperature monitoring, neuromuscular monitoring are essential.

Invasive BP monitoring can be used in patients with severe hypothyroidism undergoing major surgeries.

Possible complications

Patients may easily develop hypotension, cardiac failure, bradycardia in the post induction period and the ventilatory response to hypoxia may be decreased.

The anaesthesiologist should also be prepared for hypothermia, hyponatremia and hypoglycaemia.

Postoperative care

Reversal of neuromuscular blockade is best done with neuromuscular monitoring.

Cautious monitoring is required, anticipating hypoventilation and respiratory depression.

Postop analgesia is provided with either regional techniques (where ever possible) or non narcotic analgesics.

Information about emergency-like situations / Differential diagnostics

Myxoedema coma is a medical emergency with a reported mortality of up to 50% and needs aggressive management. It presents with hypothermia, hypoventilation hypotension, hyponatremia, and is treated with L-thyroxin at age-appropriate doses after consultation with a pediatric endocrinologist. Supportive therapy with IV fluids, thermoregulation, correction of electrolytes and cardiorespiratory support.

There might be an undiagnosed cardiac abnormality to consider in an unstable patient.

Ambulatory anaesthesia

Not reported.

Obstetrical anaesthesia

Congenital hypothyrodisim is a neonatal disease.

There is no data.

Literature and internet links

1. Klein AH, Agustin AV, Foley TP Jr. Successful laboratory screening for congenital hypothyroidism. Lancet 1974 Jul 13. 2(7872):77-9 doi:10.1016/S0140-6736(74)91637-7
2. LaFRANCHI S. Congenital Hypothyroidism: Etiologies, Diagnosis, and Management. Thyroid. July 1999, 9(7): 735-740. doi:10.1089/thy.1999.9.735
3. Medda E, Olivieri A, Stazi MA, Grandolfo ME, Fazzini C, Baserga M. Risk factors for congenital hypothyroidism: results of a population case-control study (1997-2003). Eur J Endocrinol. 2005 Dec. 153(6):765-73. Doi: 10.1530/eje.1.02048
4. Stoll C, Dott B, Alembik Y, Koehl C. Congenital anomalies associated with congenital hypothyroidism. *Ann Genet*. 1999. 42(1):17-20. PMID: 10214503
5. Parks JS, Lin M, Grosse SD, Hinton CF, Drummond-Borg M, Borgfeld L. The impact of transient hypothyroidism on the increasing rate of congenital hypothyroidism in the United States. *Pediatrics*. 2010 May. 125 Suppl 2:S54-63. doi: 10.1542/peds.2009-1975F
6. Bongers-Schokking JJ, Koot HM, Wiersma D, et al. Influence of timing and dose of thyroid hormone replacement on development in infants with congenital hypothyroidism. *J Pediatr*. 2000 Mar. 136(3):292-7. doi:10.1067/mpd.2000.103351
7. Donaldson M, Jones J. Optimising outcome in congenital hypothyroidism; current opinions on best practice in initial assessment and subsequent management. *J Clin Res Pediatr Endocrinol*. 2013 Mar 4. 5 Suppl 1:13-22. doi: 10.4274/jcrpe.849
8. Olivieri A, Stazi MA, Mastroiacovo P, Fazzini C, Medda E, Spagnolo A, De Angelis S, Grandolfo ME, Taruscio D, Cordeddu V, et al.: A population-based study on the frequency of additional congenital malformations in infants with congenital hypothyroidism: data from the Italian Registry for Congenital Hypothyroidism (1991-1998). JClinEndocrinolMetab 2002, 87(2):557-562 doi: 10.1210/jcem.87.2.8235
9. Clifton-Bligh RJ, Wentworth JM, Heinz P, Crisp MS, John R, Lazarus JH, Ludgate M, Chatterjee VK: Mutation of the gene encoding human TTF-2 associated with thyroid agenesis, cleft palate and choanal atresia. NatGenet 1998, 19(4):399-401. Doi: 10.1038/1294
10. Miller's Anesthesia, 7th Edition; Ronald D. Miller, MD, Lars I. Eriksson, Lee Fleisher, MD, Jeanine P. Wiener-Kronish, MD and William L. Young. Preoperative Evulations. p:1023
11. Kristensen MS, Moller J. Airway management behaviour, experience and knowledge among Danish anaesthesiologists – room for improvement. Acta Anaesthesiol Scand 2001; 45: 1181–1185. Doi: 10.1034/j.1399-6576.2001.450921.x
12. Bouaggad A, Nejmi SE, Bouderka MA et al. Prediction of difficult tracheal intubation in thyroid surgery. Anesth Analg 2004; 99: 603–606. Doi: 10.1213/01.ANE.0000122634.69923.67
13. Dere K, Teksoz E, Sen H, Orhan ME, Ozkan S, Dagli G.Anesthesia in a child with massive thyroid enlargement. Paediatr Anaesth. 2008 Aug;18(8):797-8. doi: 10.1111/j.1460-9592.2008.02547.x.

Last date of modification: January 2017

This guideline has been prepared by:

Author
Ahmet Eroglu, Anaesthesiologist, Karadeniz Technical University, Turkey
aheroglu@hotmail.com

Co-author
Ahmet Oguzhan Kucuk, Karadeniz Technical University, Turkey

Peer revision 1
Maurizio Delvecchio, Section of Pediatrics, Department of Biomedical Sciences and Human Oncology, University of Bari Aldo Moro, Bari, Italy
mdelvecchio75@gmail.com

Peer revision 2
James B. Bussel, Departments of Pediatrics, Medicine, and Obstetrics and Gynecology, New York Presbyterian Hospital, Weill Cornell Medical Center, New York, USA
jbussel@med.cornell.edu

Please note that this guideline has not been reviewed by two anaesthesiologists, but by two disease experts instead.