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Bullous pemphigoid

Congenital hypothyroidism

orphanaesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 7 | 2020

OrphanAnesthesia –

ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patienten mit seltenen Erkrankungen. Damit will Orphan Anesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesisten damit keine Erfahrungen gesammelt haben, so dass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

OrphanAnesthesia –

a project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

Find a survey of the recommendations published until now on:

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Anaesthesia recommendations for

Congenital hypothyroidism

Disease name: Congenital hypothyroidism

ICD 10: E03.0 with diffuse goitre
E03.1 without goitre

Synonyms: CH, neonatal hypothyroidism, foetal iodine deficiency disorder

Disease summary: 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 aetiology 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, myxoedema, goitre. In patients properly treated, there are no clinical signs.

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 aetiology 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. The 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 in 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 10-15µg/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.

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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.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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

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Typical surgery

In infants with congenital hypothyroidism, extrathyroidal 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 goitre.

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 post-operative shivering.

General anaesthesia; there are reports that thyroid surgeries performed with cervical plexus block and LMA support with spontaneous breathing. However, endotracheal intubation with flexometallic tube will be the safest option.

Necessary additional pre-operative testing (beside standard care)

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 a 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 paediatric 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 pre-medication.

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%.

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

Other problems to be anticipated are airway problems due to macroglossia, upper airway oedema, and airway deviation due to goitre.

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, transportation and mobilisation

Early mobilisation is recommended for prophylaxis of thromboembolism.

Interactions of chronic disease and anaesthesia medications

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

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

Anaesthetic procedure

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

The observed decrease in the MAC value for inhaled anaesthetics 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 cases of hypothyroidism because it will not produce hypotension and bradycardia.

Barbiturates and benzodiazepines can also be used if ketamine is contraindicated and if the 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.

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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, hyponatraemia and hypoglycaemia.

Post-operative care

Reversal of neuromuscular blockade is best done with neuromuscular monitoring.

Cautious monitoring is required, anticipating hypoventilation and respiratory depression.

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

Disease-related acute problems and effect on anaesthesia and recovery

Myxoedema coma is a medical emergency with a reported mortality of up to 50% and needs aggressive management. It presents with hypothermia, hypoventilation, hypotension, hyponatraemia, and is treated with L-thyroxin at age-appropriate doses after consultation with a paediatric endocrinologist. Supportive therapy is 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 hypothyroidism is a neonatal disease. There are no data.

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

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