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Noncompaction cardiomyopathy

Oculo-ectodermal syndrome

orphanaesthesia

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

SUPPLEMENT NR. 9 | 2023

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 Patientinnen und Patienten mit seltenen Erkrankungen. Damit will OrphanAnesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patientinnen und 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 Anästhesistinnen und Anästhesisten damit keine Erfahrungen gesammelt haben, sodass 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 eine Anästhesistin bzw. ein Anästhesist sowie eine weitere Krankheitsexpertin bzw. ein weiterer Krankheitsexperte (z. B. Pädiaterin bzw. Pädiater oder Neurologin bzw. Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, sodass 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.

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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

Noncompaction cardiomyopathy

Disease name: Noncompaction cardiomyopathy

ICD 10: 142.8 (unclassified cardiomyopathy)

Synonyms: Noncompaction cardiomyopathy. Non-compaction cardiomyopathy. Left ventricle noncompaction cardiomyopathy. Noncompaction/hypertrabeculation cardiomyopathy. Spongiform cardiomyopathy.

Disease summary: The presence of excessive and prominent trabeculae of the ventricular myocardium, together with deep intertrabecular recesses characterises the disease [1]. Three distinctive criteria define left ventricular noncompaction cardiomyopathy (LVNC): prominent left ventricular trabeculae, deep intertrabecular recesses and a thin compact layer of the myocardium [2,3]. Prevalence of the disease is uncertain. In a study with adult patients and using transthoracic echocardiography, 17/37,555 carried LVNC, 0.045 % [4], whereas in children transthoracic ultrasound showed 12/20,341 cases, 0.06 % [5]. Mortality ranges from 5 % to 47 % [6].

The disease was first recognised in the 90's of the last century as a congenital disease, due to a failure in ventricluar myocardium compaction during the 5 to 8 weeks of the embryonic development [7]. The disease was classified as an independent entity, with no age of preferential appearance, and in some cases related with other genetic disorders.

However, evidence is growing that LVNC is not a failure in the pre-existing embryonic trabecular myocardium compaction that forms the compact components of the ventricular walls [8]. When observed in adult patients, the presence of excessive trabeculae does not mean worse outcomes if the ejection fraction (EF) is normal, the risk of the development of complications, e.g. arrhythmias and stroke, being low. In fact, noncompaction images observed in children or autopsies are different from those in adult patients with excessive trabeculation, with or without clinical symptoms. Thus, it has been suggested that left-ventricle wall hypertrabeculation would not be a clinical entity by itself [8,9]. This morphological aspect could be a finding appearing together with additional lesions (as dilated cardiomyopathy) that are responsible for the low EF the patients show. The term itself can be misleading because there is neither compaction failure nor cardiomyopathy in most individuals fulfilling the diagnostic criteria.

Clinical manifestations are quite variable, ranging from asymptomatic to congestive heart failure, arrhythmias, systemic thromboembolism, and sudden death [9]. The AHA has classified the disease as a primary genetic cardiomyopathy [10], but this is controversial [11–14]. The WHO and the European Society of Cardiology classify the disease as unclassified [2], because it can be considered an independent cardiomyopathy or a phenotypical variant of other primary cardiomyopathies fulfilling the echocardiographic criteria of LVNC, i.e. dilated, hypertrophic or restrictive cardiomyopathy (with the current criteria both overlap and are not be mutually exclusive) [14]. LVNC could describe morphologic features, but not a

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functional profile of the cardiomyopathy [14,15]. There has been a shift in LVNC from underdiagnosis to overdiagnose [16].

Like other hereditary cardiomyopathies, LVNC is genetically heterogeneous [9]. LVNC1 is caused by a heterozygous mutation (autosomal dominant [17]) in the alpha-dystrobrevin gene (DTNA; 601239) in the 18q12 chromosome. However, at least 11 additional forms have been described (see Annex).

It should be taken into account that there might be a possible association between some variants and Barth syndrome or other neuromuscular disorders: dystrophinopathies, dystrobrevinopathy, laminopathy, zaspopathy, myotonic dystrophy, children glycogenosis type II (Pompe's disease), myoadenylate-deaminase deficiency, Friedreich ataxia, Duchenne's disease, Charcot-Marie-Tooth's disease and mitochondrial diseases [3,18–22].

Comprehensive reviews on LVNC are listed in references [6,15,23,24], their respective outcomes (NYHA class III or major cardiovascular complications being the worst ones, but not left ventricle dilation or systolic dysfunction) are dealt with in references [25] and [26]. Moreover, a review of paediatric cases is presented in reference [27].

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

Any type of surgery may be indicated in LVNC patients [28]. Heart failure therapy includes, in advanced forms, cardiac transplantation; thus, cardiac transplantation, extracorporeal cell membrane oxygenation (ECMO) device implant, as well as ventricular assist devices implant are relatively frequent. For the treatment of arrhythmias, automated implantable defibrillator (AID) implants can be indicated, especially in the case of ventricular arrhythmias, syncope or resuscitated sudden death. In a 6-year follow up of LVNC patients, 59 % either died or required cardiac transplant [4]. In a cohort of 113 patients who received a cardiac transplantation, outcomes were better than those whose transplant was due to ischaemic cardiopathy. However, the LVNC sample was heterogeneous, as 70 % of the patients were children [29,30].

Type of anaesthesia

There are no references comparing anaesthesia techniques or procedures. In severe cases, general anaesthesia was regularly used, but regional anaesthesia and monitored anaesthesia care (MAC) have been safely applied. Choosing one or the other will depend on the patient's haemodynamic or coagulation/anticoagulation status.

Necessary additional preoperative testing (beside standard care)

If possible, consultation with a cardiologist and haematologist is suggested.

LVNC may appear either in association with other congenital cardiomyopathies or in isolation (e.g., presenting either sporadically or familial). The age of disease presentation, evolution and the degree of affectation are variable.

Clinical manifestations are not specific. Cardiac insufficiency because of systolic or diastolic dysfunction, thromboembolic phenomena due to the combination of blood stasis in the ventricular recesses with a higher incidence of atrial fibrillation and cardiac chamber's dilatation, conduction disorders such as branch blocks, Wolf-Parkinson-White syndrome [31], supraventricular arrhythmia episodes (mainly atrial fibrillation) [32], ventricular arrhythmias [32] and sudden death may occur. Diagnostic workup consists of image techniques such as echocardiography [33], contrast echocardiography [34,35], cardiac CT and cardiac MRI [3,36–38].

Insertion of AID was reported as controversial [3] and previous Holter evaluation is suggested (outcome goals can be defined based on EF and the patient's symptoms). However, an AID is recommended if: EF is low, or EF is normal but there are a syncope history, unsustained ventricular tachycardia or a family history of cardiac sudden death [39].

Particular preparation for airway management

None described. Some genetic types can present with facial dysmorphias, leading to airway investigations.

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Particular preparation for transfusion or administration of blood products

There are no specific references. Blood products have been transfused when indicated without problems.

Particular preparation for anticoagulation

Patients having an increased risk of developing thromboembolic events are often characterised by a low EF and atrial fibrillation [3]. Oral anticoagulation could be started in patients with diagnosed ventricle thrombus or atrial fibrillation. Otherwise, a risk stratification is suggested with CHADS2/CHADS2-Vasc scores [3]. Routine anticoagulation is under debate, as thrombogenesis has not been soundly evidenced in LVNC [3].

Particular precautions for positioning, transportation and mobilisation

No special considerations.

Interactions of chronic disease and anaesthesia medications

The recommended basic treatment is identical with that of the cardiomyopathies, but evidence is low. For example, dilated cardiomyopathies with a low EF [3]. Some patients might be under beta-blocking drug therapy. A relatively distinctive treatment is anticoagulation (see above) and sudden death prevention. Regarding the latter, it needs to be considered that several neuromuscular diseases with LVNC could be related with sudden death per se [40].

Anaesthetic procedure

Sviggum et al. [28] retrospectively reviewed a cohort of 60 patients with LVNC in whom 220 surgical procedures had been performed. Nineteen patients had suffered from 25 complications, 10 being new arrhythmias, 5 respiratory, one seizure and one syncope. 47 % of these complications had occurred during open cardiac bypass procedures under general anaesthesia, and none with regional anaesthesia or monitored anaesthesia care/sedation. There were no cases of long-term morbidity or perioperative mortality in this series. The authors pointed out that complication rates were not different from those without LVNC. In some cases, the disease was diagnosed after a severe heart insufficiency workup for cardiac transplantation [35].

A case of cardiac arrest during the induction of sevoflurane anaesthesia for a dental procedure in a child with LVNC, with complete recovery, has been reported [41].

A few surgical cases have been published. A young male patient had suffered a traumatic spleen rupture [42]. There had been a history of severe cardiopathies in his family. Six months before, the patient had complained of congestive heart failure with systolic ventricular dysfunction and paroxysmal atrial fibrillation, and LVNC was diagnosed. An AID was inserted and acenocoumarol anticoagulation started. Before surgery (open splenectomy), the AID was disconnected and transcutaneous pacing pads applied. General anaesthesia consisted

of midazolam 0.1 mg/kg, ketamine 100 mg, rocuronium 50 mg, and 50 % O_2/air, sevoflurane and rocuronium infusion maintenance, as well as fentanyl boluses. The patient was transfused red packed cells, fresh frozen plasma and platelets. 48 hours after ICU admission, oral anticoagulation was restarted (60 mg/24 h of enoxaparin have been administered till this moment previously).

Kim et al. [43] published the case of a female patient under laparoscopic ovarian cystectomy. LVNC had been diagnosed before. The patient showed sinus bradycardia and a 1st degree auriculo-ventricular block, multiple premature ventricular contractions and a left branch block. Severe left ventricle dysfunction with an EF of 30 % and a left atrial dilatation were also observed. Invasive arterial pressure and transoesophageal echocardiography (TEE) were used for monitoring. General anaesthesia with etomidate, midazolam, cis-atracurium and propofol-remifentanil was selected. A dobutamine infusion was needed after induction. No other alterations were observed during the procedure including CO_2 insufflation periods. During the 24 h ICU admission there were no incidences.

Kumar et al. [44] reported the case of a patient with biventricular noncompaction cardiomyopathy with Ebstein anomaly and a mass in the left atrium. An external assist device was inserted into the ventricle. Due to the thin wall of the ventricle and trabeculae, the inflow cannula was correctly inserted thanks to TEE. The patient's heart was transplanted afterwards.

There have been cases of malignant hyperthermia among LVNC patients possibly coinciding with neuromuscular diseases. Two cases reportedly occurred intraoperatively, one during the insertion of a biventricular assist device (after ECMO) in a 25-year-old patient who had sustained a refractory cardiogenic shock [45], the other during cardiac surgery [46].

Particular or additional monitoring

During the perioperative management of LVNC patients, preoperative evaluation is fundamental to know the haemodynamic status and select the surgical and anaesthesia techniques, as well as monitoring and perioperative patient care, in order to diminish risks.

In the described haemodynamically unstable trauma patient, with LVNC, monitoring consisted in ECG (DII derivation) to check for rhythm alterations, urine output control, and invasive arterial pressure. Several months before the patient had complained of atrial fibrillation with a rapid ventricular response and acute cardiac insufficiency that led to the diagnosis of cardiomyopathy [42]. Due to the arrhythmogenesis, ECG monitoring and the rapid treatment of arrhythmias was recommended. In LVNC patients with cardiac insufficiency, pulmonary artery catheter insertion or TEE have been used [43,47,48] to determine preload and ventricular function. In the case reported, the patient carried an AID requiring disconnection and external pacing pads sited perioperatively [42]. Specific information about AID indications can are mentioned in reference [49]. Management (general) of AID is shown in table 1.

Possible complications

Cardiac insufficiency, arrhythmias, systemic thromboembolism, haemorrhage, sudden death.

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Postoperative care

Depending on the surgery and on their previous haemodynamic status, patients with LVNC might need ICU admission. ICU admission is suggested in moderate to severe cases, and in those with actual or foreseen haemodynamic instability. In most of the published cases, patients were admitted to an ICU because of severe complications. In one published case, an episode of ventricular fibrillation was registered and treated within the first postoperative hours in the ICU as the AID was adequately restarted [41].

Anticoagulation should be reintroduced as soon as possible to prevent thromboembolic events (both if the patient is under anticoagulation therapy or when risk factors for thrombus formation concur).

Disease-related acute problems and effect on anaesthesia and recovery

There are no specific recommendations. A case-by-case evaluation is mandatory, due to the clinical variability.

Ambulatory anaesthesia

There are no specific references published.

Obstetrical anaesthesia

Several obstetrical cases have been communicated. As previously stated in other settings, clinical presentation and evolution of LVNC during pregnancy is variable [49]. In a previously diagnosed LVNC patient who suffered severe symptoms and was scheduled for cesarean section under general anaesthesia, invasive monitoring was started including pulmonary artery catheterisation with pacing, as well as entry ports for arteriovenous ECMO. Dobutamine and milrinone infusions were also applied. Anaesthesia induction was performed with S-ketamine 0.5 mg/kg, etomidate 0.25 mg/kg and succinylcholine. Maintenance was done with propofol/remifentanil. The patient's course was stable. She was admitted to the ICU with no incidences and required no ECMO [50]. Another patient with preterm gestation and preeclampsia, sustaining severe LVNC and pulmonary hypertension, needed a cesarean section. Monitoring included TEE instead of a pulmonary artery catheter [48]. In another cesarean section case, a patient with severe LVNC developed a postpartum haemorrhage after being unresponsive to several treatments for uterine atonia. In this patient, pulmonary hypertension and severe right ventricular insufficiency developed immediately after intramuscular methylergonovine injection. Inotropic drug support was needed, and she was admitted to the ICU with good evolution [51]. Uesugi et al. [52] reported of a 24 weeks pregnant patient who was scheduled for cesarean section. However, symptomatic cardiac failure developed. Anaesthesia consisted of propofol and fentanyl, with intraoperative haemodynamic stability. Two years later, she needed another cesarean section that was performed under spinal anaesthesia in her 34th gestational week, because she had normal cardiac function and no anticoagulation. No incidences were observed.

In other reported cases, there was no previous LVNC diagnosis. In a pregnant patient, a cerebral infarction due to embolism of cardiac origin was attributed to LVNC. Several days after stabilisation, an elective cesarean section was performed under general anaesthesia

[53]. In a pregnant woman with systolic failure of the left ventricle, a condition which might develop during labour, general anaesthesia consisted of etomidate, midazolam, succinylcholine and fentanyl-midazolam boluses. Invasive monitoring with arterial pressure, central venous catheter and TEE that led to the diagnosis of LVNC. A dobutamine infusion was needed and no further incidences were reported after ICU admission. In the postoperative period, angiotensin-converting enzyme inhibitors and beta-blocking drugs were administered, also oral anticoagulation and an AID [54]. Finally, a 25-year-old woman suffered a dilated cardiomyopathy and recovered from cardiac arrest 8 weeks after labour. TEE during anesthesia to insert a left ventricular assist device revealed apical LVNC. Authors comment that this is the first reported case of such a combination (peripartum-dilated cardiomyopathy and LVNC) [55]. However, as stated above, the development of another cardiomyopathy can be taken into account.

Table 1: General management of IAD.

Pacemaker (PM)-dependent patient or device in rate-dependent mode:
YES.
Reprogram device with proprietary programmer: (1) Inactivate rate response or program to asynchronous pacing mode if PM-dependent AND (2) suspend anti-tachycardia therapy.
NO.
IAD accessible: Place magnet over AID, perform surgery, remove magnet.
IAD no accessible: (1) AID not accessible OR (2) magnet not securely applicable OR uncertain magnet response (audio/vibrate/pacing): reprogram device with proprietary programmer to suspend anti-tachycardia therapy.

Reference: Sticherling C, Menafoglio A, Burri H, Reek S, Fuhrer J, Ganière V, et al. Recommendations for the peri- operative management of patients with cardiac implantable electronic devices. Med Cardiovasc 2016;19:13–18.

Annex.

Genetic heterogeneity of LVNC12:

LVNC1, heterozygous mutation of the alpha-dystrobrevin gene (DTNA; 601239), 18q12 chromosome; locus of an autosomal dominant form, 11p15 chromosome (LVNC2; 609470); LVNC3 (see 605906), mutation in LDB3 gene (605906), 10q23 chromosome; LVNC4 (see 613424) mutation in ACTC1 gene (102540), 15q14 chromosome; LVNC5 (see 613426) mutation in MYH7 gene (160760), 14q12 chromosome; LVNC6 (see 601494) mutation in TNNT2 gene (191045), 1q32 chromosome; LVNC7 (615092) mutation in MIB1 gene (608677), 18q11 chromosome; LVNC8 (615373) mutation in PRDM16 gene (605557), 1p36 chromosome; LVNC9 (see 611878) mutation in TPM1 gene (191010), 15q22 chromosome; LVNC10 (615396) mutation in MYBPC3 gene (600958) 11p11 chromosome; LVNC can take part of a X-linked disorder, Barth syndrome (302060), caused by a mutation in TAZ gene (300394), Xq28 chromosome.

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