orphanaesthesia

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 ventricle noncompaction cardiomyopathy (LVNC): prominent left ventricle trabeculae, deep intertrabecular recesses and a thin compact layer of the myocardium [2,3]. Prevalence of the disease in uncertain. In a study in adult patients 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 5 % to 47 % [6].

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

However, evidence is growing that LVNC is not a failure in the pre-existing embrionic trabecular myocardium compaction that forms the compact components of the ventricle walls [8]. When observed in adult patients, the presence of excessive trabeculae does not comport worse outcomes if the ejection fraction (EF) is normal, the risk of the development of complications, as 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 of 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 echocardiography criteria of LVNC, as dilated, hypertrophic or restrictive cardiomyopathy (with the current criteria it can overlap and not be mutually exclusive) [14]. LVNC could describe morphologic features but not a functional profile

of the cardiomyopathy [14,15]. There has been a passage in LVNC from infra to overdiagnose [16].

As other hereditary cardiomyopathies, LVNC is genetically heterogeneous [9]. LVNC1 is caused by a heterozigous mutation (autosomal dominant17) 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 the possible association of some variants with Barth syndrome and with other neuromuscular disorders: dystrophinopathies, dystrobrevinopathy, lamino-pathy, zaspopathy, myotonic dystrophy, children glucogenosis type II (Pompe's disease), myoadenilate-deaminase deficiency, Friedreich ataxia, Duchenne's disease, Charcot-Marie-Tooth's disease and mitochondrial diseases [3,18–22].

Thorough reviews of LVNC can be read in references [6,15,23,24] and outcomes (NYHA class III or major cardiovascular complications as the worse ones, but not left ventricle dilation or systolic dysfunction) 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

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Typical surgery

Any type of surgery can 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 arrhythmia's treatment 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 % were dead 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 patients were children [29, 30].

Type of anaesthesia

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

Necessary additional pre-operative testing (beside standard care)

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

LVNC can be associated to other congenital cardiomyopathies, or appear isolated (in this case being sporadic or familial). Age of disease presentation, evolution, as well as 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 as branch blocks, Wolf-Parkinson-White syndrome [31], supraventricular arrhythmia episodes (mainly atrial fibrillation) [32], ventricular arrhythmias [32] and sudden death can occur. Diagnostic workup consist in image techniques such as echocardiography [33], contrast echocardiography [34,35], cardiac CT and cardiac MRI [3,36–38].

Insertion of AID was reported as controversial3 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 dysmorphies, leading to airway investigations.

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 increased risk to develop thromboembolic events are these with 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 that of the cardiomyopathies, but evidence is low. As an example, those of dilated cardiomyopathies with low EF [3]. Some patients can be under beta-blocking drug therapy. A relatively distinctive treatment is anticoagulation (see before) and sudden death prevention. Regarding the latter, it need 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 revised a cohort of 60 patients with LVNC in whom 220 surgical procedures were performed. Nineteen patients suffered of 25 complications, 10 being new arrhythmias, 5 respiratory, one seizure and one syncope. 47 % of these occurred during open cardiac bypass procedures under general anaesthesia, and none with regional anaesthesia or monitored anaesthesia care/sedation. There were neither long-term morbidity nor peri-operative mortality in this series. Authors pointed out that complication rates were not different from those without LVNC. In some cases, the disease was diagnosed after severe heart insufficiency workup for cardiac transplantation [35].

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

There are few published surgical cases. A young male patient suffered a traumatic spleen rupture42. A family history of severe cardiopathies was reported. Six months ago, the patient complained from congestive heart failure with systolic ventricular dysfunction and paroxismal atrial fibrillation, and LVNC was diagnosed. An AID was inserted and acenocumarol 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 % O2/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).

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 1st degree auriculo-ventricular block, multiple premature ventricular contractions and left branch block. Severe left ventricle dysfunction with EF 30 % and left atrial dilatation were observed too. Invasive arterial pressure and transoesophageal echocardiography (TEE) were used for monitoring. General anaesthesia with etomidate, midazolam, cis-atracurium and propofol-remifentanil was selected. Dobutamine infusion was needed after induction. No other alterations were observed during the procedure including CO2 insufflation periods. During the 24h 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 in 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.

Malignant hyperthermia cases have been described in LVNC patients in possible relationship with the coincidence with neuromuscular diseases. Two cases have been reported intraoperatively, one during a biventricular assist device insertion (after ECMO) in a 25 year-old patient sustaining refractory cardiogenic shock [45], and 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 to 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. The patient complained several months before of atrial fibrillation with rapid ventricular response and acute cardiac insufficiency that lead to the diagnosis of the cardiomyopathy [42]. Due to the arrhythmogenesis, ECG monitoring is recommended, as well as rapid treatment of arrhythmias. 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 peri-operatively [42]. Specific information about AID indications can be read in reference [49]. Management (general) of AID is showed in table 1.

Possible complications

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

Post-operative care

Depending on the surgery and on the previous haemodynamic status, the patient with LVNC might need ICU admission. UCI 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 a published case, a ventricular fibrillation episode was registered and treated in 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 in the case the patient was under anticoagulation therapy or when risk factors for thrombus formation concurred).

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

There are several obstetrical cases 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 who was scheduled for cesarean section under general anaesthesia, invasive monitoring was started that included pulmonary artery catheterisation with pacing, as well as entry ports for arteriovenous ECMO. Dobutamine and milrinone infusions were started, too. Anaesthesia induction consisted in S-ketamine 0.5 mg/kg, etomidate 0.25 mg/kg and succinylcholine. Maintenance was with propofol/remifentanil. The patient course was stable. She was admitted to the ICU with no incidences and with no ECMO use needed [50]. Another patient with preterm gestation and preeclampsia, sustaining severe LVNC and pulmonary hypertension, needed a cesarean section. Monitoring included TEE instead of pulmonary artery catheter [48]. In another cesarean section case, a patient with severe LVNC developed 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 methyl-ergonovine injection. Inotropic drug support was needed, and she was admitted to the ICU with good evolution [51]. Uesugi et al. [52] reported on 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 afterwards, 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 were 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 that develops during labour, general anaesthesia consisted of etomidate, midazolam, succinylcholine and fentanyl-midazolam boluses. Invasive monitoring was with arterial pressure, central venous catheter and TEE that lead to LVNC diagnosis. Dobutamine infusion was needed, and after ICU

admission, no more incidences were reported. In the postoperative period, angiotensin-converting enzyme inhibitors and beta-blocking drugs were started, as was 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 case reporting such a combination (peripartum dilated cardiomyopathy and LVNC) [55]. However, as stated before, the development as a secondary cardiomyopathy cannot be ruled out.

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.

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Annex.

Genetic heterogeneity of LVNC12:

LVNC1, heterozigotic mutation of alpha-dystrobrevin gene (DTNA; 601239), 18q12 chromosome; locus for 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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Date last modified: **December 2021**

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Disclosure The author has no financial or other competing interest to disclose. This recommendation was unfunded.

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Disclosure The reviewers have no financial or other competing interest to disclose.