www.cardiologyrounds.ca 2008 volume XIII, issue 3

CARDIOLOGY Rounds®

As presented in the rounds of the Division of Cardiology, St. Michael's Hospital, University of Toronto

Isolated Ventricular Noncompaction: A New Cardiomyopathy

By PAALADINESH THAVENDIRANATHAN, MD, MSc, FRCPC, and CHI-MING CHOW, MDCM, MSc, FRCPC, FACC

Isolated ventricular noncompaction (IVNC) is a rare congenital disorder that was first described in a 33-year-old woman by Engberding et al,[1] in 1984. Subsequently, many case reports and several case series in both children and adults have been published.[2-9] IVNC was recently classified as a genetic cardiomyopathy[10] caused by a failure of normal embryogenesis of the endocardium and myocardium *in utero*,[11] resulting in prominent ventricular trabeculations with deep intertrabecular recesses (Figure 1). Noncompaction can occur in isolation or in association with various other cardiac anomalies, such as obstruction of the right or left ventricular outflow tract, complex cyanotic congenital heart disease, and coronary anomalies.[12,13] However, IVNC, by definition, occurs in the absence of other coexisting cardiac abnormalities.[14]

The clinical manifestations of this disease are quite diverse and often diagnosis is delayed due to the limited awareness of its unique clinical and imaging characteristics.[5] This issue of *Cardiology Rounds* summarizes the literature on IVNC with respect to embryogenesis, clinical presentation, diagnostic criteria, management, and prognosis.

Epidemiology

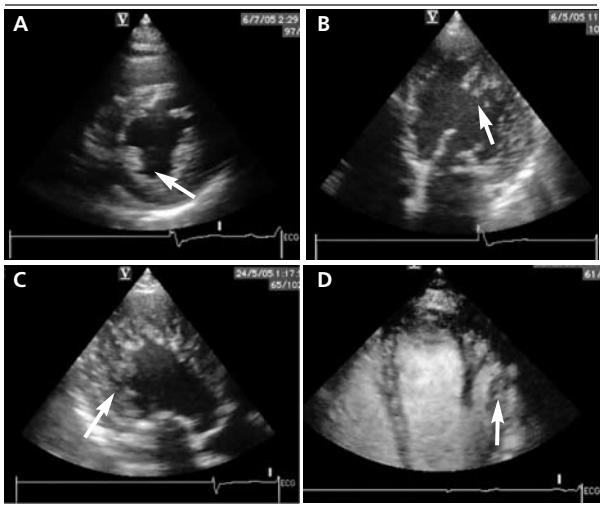
IVNC has been described both in children and in adults. Based on all current case reports, the incidence is higher in men than in women. A recent epidemiological study of primary cardiomyopathies in Australian children indicated that IVNC accounted for 9.2% of all cases and that it was the third most common cause of cardiomyopathy after dilated and hypertrophic cardiomyopathies.[15] This is similar to the experience at the Texas Children's Hospital in the United States (US), where IVNC accounted for 9.5% of cardiomyopathies identified in children over a 5-year period.[9] In the adult population, among patients referred to a tertiary-care centre for echocardiography, two series of case reports indicated a prevalence of 1.4 to 26 per 10,000;[4,16] nevertheless, since all current prevalence data are based on case reports, the true prevalence of IVNC in adults remains unclear.

Genetics

IVNC is a genetically heterogeneous disorder with both familial and sporadic forms. To date, mutations in 7 different genes have been found;[17] however, at present, mutations in G4.5 gene (TAZ gene) on the Xq28 chromosomal region identified in neonatal IVNC is the only confirmed disease-causing locus.[18] This gene codes for a protein known as taffazin, which has a role in the maintenance of the cardiolipin layer in the inner membrane of the mitochondria, as well as in the promotion of differentiation and maturation of osteoblasts, and in prevention of maturation in adipocytes.[19] Mutations in the TAZ gene have not been identified in adults; further, contrary to the potential X-linked transmission in neonatal IVNC, it has been proposed that the mechanism of transmission in adults is autosomal dominant.[20] This is based on the observations that approximately 50% of descendants of patients with IVNC inherit the condition, cases of male-to-male transmission occur, and the disorder may occur in females.

St. Michael's Hospital
30 Bond St.,
Suite 7049, Queen Wing
Toronto, Ont. M5B 1W8
Fax: (416) 864-5941

Figure 1: Echocardiographic views of the left ventricle with and without contrast illustrating noncompaction



A-C: Noncompaction in the short axis, apical four-chamber, and apical three-chamber views, respectively.
D: Contrast echo imaging showing the areas of noncompaction in the left-ventricular lateral wall.

Cardiac embryology

A thorough understanding of IVNC necessitates knowledge of cardiac embryogenesis. During weeks 1-2 of life *in utero*, cardiac myocytes migrate and form the primitive endocardium and myocardium.[21] At 3 weeks, the 2 layers fuse into a single beating cardiac tube with no epicardial overlay.[21] In an adaptation to improve the nourishment for a rapidly growing heart, the myocardium forms a loose network of interwoven fibres (trabeculations) that is connected to the left ventricular (LV) cavity via deep recesses (Figure 2).[21] The trabeculations increase the surface area and facilitate myocardial nutrition supply by exchange diffusion. By weeks 5-8, an epicardial layer forms around the myocardium and a vascular network begins to form in this subepicardial space, eventually becoming the predominant source of myocardial nutrition.[21] During this period, the trabeculated myocardium compacts, gradually proceeding from the epicardium to the endocardium and from the base to the apex in the same direction as the formation of epicardial coronary vessels.[5] The intertrabecular recesses are converted into capillaries that then connect to the epicardial vessels.[22] This process of compaction is usually completed in the early fetal period, resulting in a prominent outer compact myocardium with a few residual subendocardial trabeculae (Figure 2).

Ventricular noncompaction is thought to occur due to an interruption in this normal process of myocardial morphogenesis.[21] The time of arrest of this process will determine the extent of myocardial noncompaction within the ventricle;[21] however, the reason for a premature arrest in this process is not well understood. There is evidence using magnetic resonance imaging (MRI), positron emission tomography (PET), and ^{201}thallium scintigraphy that

Figure 2: Scanning electron microscopy pictures illustrating the process of normal compaction during embryological life

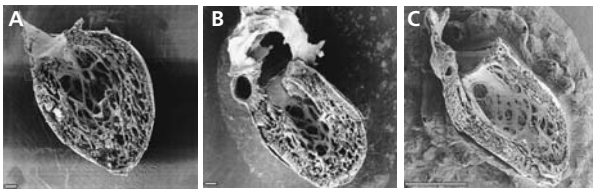


Figure 2A: Reproduced from Sedmera D et al.[22] Copyright © 2000 Wiley-Liss Inc. Reprinted with permission.
Figures 2B and 2C: Reproduced from Wessels A et al.[23] Copyright © 2003 the American Physiological Society. Reprinted with permission.

disturbances in microcirculation are present in the hearts of patients with IVNC corresponding to the areas of noncompaction.[21] Furthermore, coronary flow reserve is reduced in noncompacted segments as well as in other segments with wall motion abnormalities.[21] However, it is unclear whether the persistence of an embryonic pattern of trabeculated myocardium is secondary to a growth failure of coronary microcirculation within the increased ventricular mass or whether the abnormal myocardial development prevented the normal progression of microvascular coronary artery development.[21]

Clinical presentation

The clinical presentation of IVNC is highly variable, ranging from an incidental echocardiographic diagnosis without symptoms to disabling heart failure. Often, the reasons for referral to a cardiologist in patients eventually diagnosed with IVNC include unexplained heart failure, uncertain echocardiographic findings, palpitations, or shortness of breath on exertion.[4] The 3 most common clinical presentations include heart failure (HF), arrhythmias, and embolic events;[11,24] additional presenting symptoms include chest pain and syncope.[24]

Heart failure

In HF, clinical presentations range from asymptomatic LV dysfunction to severe disabling HF. In a comparison of all published adult case series, the prevalence of symptomatic HF is 30%-73%, while LV systolic dysfunction is 58%-82%.[2-9] This is similar to the prevalence in case reports of children with IVNC. Although most patients have systolic LV dysfunction, diastolic dysfunction has also been described in as many as one-third of the patients in several case series.[3,24,25] The cause of systolic dysfunction in patients with IVNC is unclear; however, there are two leading hypotheses:

- chronic subendocardial hypoperfusion and microcirculatory dysfunction[26]
- reduced thickness of the compact myocardial layer in relation to the trabeculated myocardium in the affected regions.[21]

Diastolic dysfunction, on the other hand, may be related to both abnormal relaxation and restrictive filling caused by the numerous prominent trabeculae.[25]

Arrhythmias

The 2 most common, clinically significant arrhythmias in patients with IVNC are atrial fibrillation and ventricular tachycardia. Atrial fibrillation is seen in as many as 5%-29% of patients, while ventricular tachycardia has been described in 3%-47% of patients.[2-9] Sudden death occurred in 18% of patients followed in 2 case series.[4,5] Paroxysmal supraventricular tachycardia and complete heart block have also been reported in patients with IVNC.[3,5] Several hypotheses have been proposed for the high incidence of ventricular arrhythmias and sudden cardiac death in patients with IVNC. These include irregular branching and connection of myocardial fascicles to the noncompacted segments, isometric contraction with increased wall stress, and localized coronary perfusion impairment inducing disorganized or delayed electrical activation.[27] In addition, postmortem analyses have illustrated the presence of ischemic subendocardial lesions with fibrosis, likely secondary to chronic ischemia. These lesions can also act as substrates for ventricular arrhythmias;[28] furthermore, many patients with IVNC have significant LV dysfunction, which in turn is a risk factor for ventricular arrhythmias.[24]

Several nonspecific electrocardiogram (ECG) abnormalities are present in most patients with IVNC;[8,24] these include LV hypertrophy, left bundle branch block, intraventricular conduction delays, inverted T waves, and axis shifts. In approximately 15% of pediatric patients, Wolff-Parkinson-White syndrome has been described; however, its prevalence in the adult population is negligible.

Thromboembolism

Thromboembolism may include the cerebrovascular, pulmonary, peripheral vascular, or mesenteric systems. Initial case reports illustrated a prevalence as high as 24% in adult populations;[5] however, more recent publications indicate a prevalence of 0%-9% over a follow-up period of 2.4–3.8 years.[7,8,24] This lower prevalence is partially due to selective anticoagulation with acetylsalicylic acid or warfarin in 2 of the 3 recent case reports and the larger populations reflect a more accurate estimate.[7,24] The prevalence of thromboembolism in the 2 pediatric case series is much lower and no cases of systemic thromboembolism are reported in the largest case series. Thromboembolic events have been attributed to the formation of thrombi in the extensively trabeculated ventricular myocardium, to LV dysfunction, and/or atrial fibrillation. Nevertheless, recent pathological data suggest that the formation of thrombi in noncompacted ventricles is not very common.[29] Furthermore, one of the largest case-control retrospective studies in patients with IVNC revealed that when patients were matched with controls having similar degrees of LV dysfunction, there was no difference in the rates of systemic thromboembolism.[30] As a result, it appears that the main risk factors for cardioembolic events in patients with IVNC are the severity of the underlying systolic dysfunction, the presence of atrial tachyarrhythmias, and the presence of previous thromboembolic events rather than noncompaction itself.

Associated features

Concomitant with IVNC, in pediatric case series, facial dysmorphic features (eg, prominent forehead, strabismus, low-set ears, high-arched palate, and micrognathia) have been described;[3] however, these features have not been observed in adults.[5,8] Instead, in the adult population, Stollberger et al[6] described a high incidence of neuromuscular disorders (NMD) in association with IVNC. Although the prevalence was much lower, this finding is supported in a case series by Lofiego et al,[7] but it is not found in 2 other case series.[8,24]

Diagnosis

Echocardiography

Traditionally, the diagnosis of IVNC is based on 2D echocardiography and characterized by the presence of numerous, prominent trabeculations with deep intertrabecular recesses in hypertrophied and often hypokinetic segments in the myocardium of the LV (Figure 1).[11] The most commonly involved areas are the LV apex, and the mid inferior and lateral walls.[28] Ventricular-wall hypokinesis may also be observed in the compacted segments of the myocardium, which supports the hypothesis of diffuse myocardial microcirculatory dysfunction in these patients.[4] The most commonly used echocardiographic criteria for the diagnosis of IVNC in adults follows the proposal by Oechslin et al (Table 1, Figure 3).[14] The validity of these criteria in differentiating IVNC from other common cardiomyopathies was established in a subsequent publication.[31]

Right-ventricular noncompaction has also been described; however, due to the difficulty in distinguishing normal variants found in the highly trabeculated right ventricle from the pathological noncompacted ventricle, many authors dispute the existence of this entity.[4] Currently, standard criteria for the diagnosis of right-ventricular noncompaction do not exist.

Magnetic resonance imaging

MRI has an emerging role in the diagnosis and prognosis of patients with IVNC.[32] Currently, the role of MRI includes confirming echocardiographic findings, detecting subtle forms of IVNC, localizing and determining the extent of myocardial involvement, obtaining myocardial perfusion data, assessing extent of myocardial fibrosis, identifying ventricular thrombi, and helping to differentiate IVNC from other potential diagnoses.[33] MRI findings for IVNC reveal (Figure 4):

- numerous, excessive trabeculations in the LV with predominant involvement of apical and mid portions of the lateral and inferior walls[32]
- thinning of the LV wall in diastole0

Table 1: Diagnostic criteria for IVNC of the myocardium by Oechslin et al[4]

1. Absence of coexisting cardiac abnormalities (other than 2-4 below) by definition
2. Typical 2-layered structure of the myocardium with a thin, compacted outer (epicardial) band and a much thicker, noncompacted inner (endocardial) layer consisting of trabecular meshwork with deep endocardial spaces (the maximum end systolic of the noncompacted to compacted myocardium of >2 is characteristic). Measure in parasternal short axis at end systole (Figure 3).
3. Predominant segmental location of the abnormality (ie, noncompacted myocardium is predominantly [<80%] found in the apical and midventricular areas of both the inferior and lateral wall)
4. Colour Doppler echocardiographic evidence of deeply perfused intertrabecular recesses (without communication with coronary circulation)

- presence of a 2-layered myocardium with non-compacted to compacted myocardial ratio at end-diastole >2.3.[33]

Furthermore, several MRI features are thought to have poor prognostic value; these include high intensity T2 signal in the endocardial layer,[34] presence of subendocardial perfusion defects,[35] and delayed enhancement of the subendocardial layer.[36]

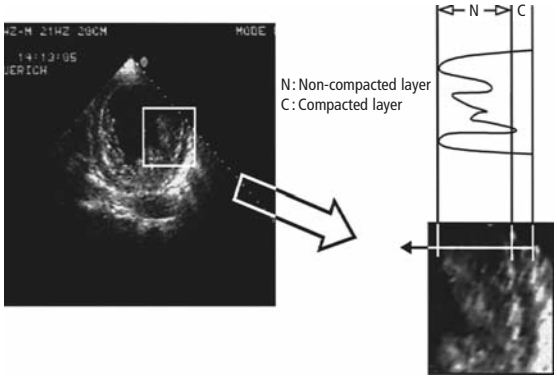
Other diagnostic modalities

Several other diagnostic modalities have been described; however, none have proven superior to echocardiography or MRI. To date, computed tomography (CT) has not been widely used in the description of patients with IVNC and no diagnostic criteria exist. PET has been used to illustrate the presence of microcirculatory dysfunction in the hearts of patients with IVNC,[26] but it lacks utility in the diagnosis of IVNC. The "loosened myocardium" of IVNC has also been observed by ventriculography during angiography.[37] However, contrast ventriculography and coronary angiography are useful to rule out other concomitant cardiac abnormalities, rather than to diagnose IVNC.

Natural history

Prognosis for patients with IVNC is highly variable; the spectrum ranges from a prolonged asymptomatic course to rapid progressive HF, which may lead to heart transplantation or death. Based on a pediatric case series, LV dysfunction inevitably develops in patients with IVNC over a 10-year period, regardless of the presence of symptoms at the time of diagnosis.[3] Similarly, in an adult case series with serial echocardiograms, 43% of patients had a progressive decline

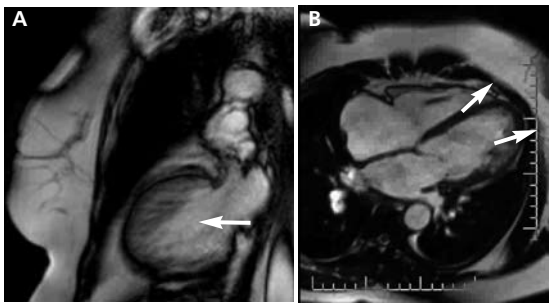
Figure 3: Short-axis view illustrating compacted (C) and noncompacted (N) areas in a patient with IVNC. Ratio of N/C >2 is included in the Oechslin et al[4] criteria



Reproduced from Jenni R et al. *Heart.* 2001;86(6):666-671.[14] Copyright © 2001, BMJ Publishing Group Ltd. and the British Cardiac Society. Used with permission.

in LV ejection fraction over a 3-year period.[24] HF hospitalization is common in adults, occurring in as many as 50% of patients in one study.[4,7,24] Earlier studies by Ritter et al[5] and Oechslin et al[4] reported mortality in the range of 35%-47% for mean follow-up periods of 2.5 and 3.7 years, respectively. More recent publications with similar mean follow-up periods report mortality in the range of 2%-15%, suggesting that the prognosis may not be as poor as previously described. This is likely because many patients identified in recent studies had milder phenotypes of the disease, lower incidences of symptoms at diagnosis, higher prevalences of implantable cardioverter defibrillator (ICD) use, better use of evidence-based medical therapy, and improved echocardiographic techniques to facilitate the detection of previously unrecognized asymptomatic cases. Certain features seem to be associated with patients at a higher risk of mortality, including higher LV end-diastolic diameter at the time of initial presentation,

Figure 4: MRI images illustrating left-ventricular noncompaction



A: MRI image showing area of noncompacted myocardium (arrow). **B:** Illustration of noncompaction in the lateral left ventricular wall (lower arrow), and thinning of the left ventricle in diastole (upper arrow).

New York Heart Association class III/IV, chronic atrial fibrillation, bundle branch block,[4] sustained ventricular arrhythmias, and higher left-atrial size.[7] Patients with these characteristics may need closer follow-up and more aggressive clinical management.

Management

Currently, there are no guidelines for the management of patients with IVNC. Management plans generally involve a confirmation of the diagnosis with echocardiogram and other imaging modalities, as needed. Prominent trabeculations (normal variant), apical hypertrophic cardiomyopathy, hypertensive heart disease, dilated cardiomyopathy, endocardial fibroelastosis, cardiac metastases, and LV thrombus are important differential diagnoses.[3,6,24] Patients with noncompaction with or without LV systolic dysfunction should be followed by a cardiologist on a regular basis, with the frequency based on symptoms. Clinical visits should comprise a history, physical examination, and echocardiography, as well as Holter monitoring to identify silent arrhythmias.[5,7,27] Symptomatic patients should be managed based on their clinical presentation, following the respective consensus guidelines.[7,23] There has been significant controversy regarding anticoagulation for patients with IVNC. Some argue that all patients should be anticoagulated with warfarin,[4,5] while others recommend anticoagulation for those with LV dysfunction, atrial fibrillation, a previous history of embolic events, or those with known ventricular thrombi.[29] The issue of ICD implantation in these patients because of the high risk of sudden cardiac death is also highly controversial.[4] Some authors propose that patients with high-risk predictors of death (see above) should be considered for ICD implantation.[27] Finally, first-degree family members of all patients diagnosed with IVNC should undergo a screening echocardiogram.[4]

Conclusion

IVNC is a rare genetic congenital disorder caused by failure of the normal embryogenic myocardial compaction process. Its clinical presentation is highly variable, ranging from incidental echocardiographic diagnosis to symptomatic heart failure, thromboembolism, or arrhythmia. The main diagnostic modalities are 2D echocardiogram and MRI, both of which have clearly defined diagnostic criteria. Patients with IVNC are at risk for many adverse clinical outcomes and need to be followed closely by clinicians with adequate familiarity with the disease. Management is based on guidelines that have been set out for the various clinical presentations, and screening of first-degree relatives is warranted.

Dr. Thavendiranathan *is a trainee in the Adult Cardiology Training Program at the University of Toronto.*

Dr. Chow *is Staff Cardiologist, St. Michael's Hospital, and Assistant Professor, University of Toronto.*

References

1. Engberding R, Bender F. Identification of a rare congenital anomaly of the myocardium by two-dimensional echocardiography: persistence of isolated myocardial sinusoids. *Am J Cardiol.* 1984;53(11):1733-1734.
2. Chin TK, Perloff JK, Williams RG, Jue K, Mohrmann R. Isolated noncompaction of left ventricular myocardium. A study of eight cases. *Circulation.* 1990;82(2):507-513.
3. Ichida F, Hamamichi Y, Miyawaki T, et al. Clinical features of isolated noncompaction of the ventricular myocardium: long-term clinical course, hemodynamic properties, and genetic background. *J Am Coll Cardiol.* 1999;34(1):233-240.
4. Oechslin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R. Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. *J Am Coll Cardiol.* 2000;36(2):493-500.
5. Ritter M, Oechslin E, Sütsch G, Attenhofer C, Schneider J, Jenni R. Isolated noncompaction of the myocardium in adults. *Mayo Clin Proc.* 1997;72(1):26-31.
6. Stollberger C, Finsterer J, Blazek G. Left ventricular hypertrabeculation/noncompaction and association with additional cardiac abnormalities and neuromuscular disorders. *Am J Cardiol.* 2002;90(8):899-902.
7. Lofiego C, Biagini E, Pasquale F, et al. Wide spectrum of presentation and variable outcomes of isolated left ventricular non-compaction. *Heart.* 2007;93(1):65-71.
8. Murphy RT, Thaman R, Blanes JG, et al. Natural history and familial characteristics of isolated left ventricular non-compaction. *Eur Heart J.* 2005;26(2):187-192.
9. Pignatelli RH, McMahon CJ, Dreyer WJ, et al. Clinical characterization of left ventricular noncompaction in children: a relatively common form of cardiomyopathy. *Circulation.* 2003;108(21):2672-2678.
10. Maron BJ, Towbin JA, Thiene G, et al. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. *Circulation.* 2006;113(14):1807-1816.
11. Jenni R, Oechslin EN, van der Loo B. Isolated ventricular noncompaction of the myocardium in adults. *Heart.* 2007;93(1):11-15.
12. Dusek J, Ostadal B, Duskova M. Postnatal persistence of spongy myocardium with embryonic blood supply. *Arch Pathol.* 1975;99(6):312-317.
13. Lauer RM, Fink HP, Petry EL, Dunn MI, Diehl AM. Angiographic demonstration of intramyocardial sinusoids in pulmonary-valve atresia with intact ventricular septum and hypoplastic right ventricle. *N Engl J Med.* 1964;271:68-72.
14. Jenni R, Oechslin E, Schneider J, Attenhofer Jost C, Kaufmann PA. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. *Heart.* 2001:86(6):666-671.
15. Nugent AW, Daubeney PE, Chondros P, et al; National Australian Childhood Cardiomyopathy Study. The epidemiology of childhood cardiomyopathy in Australia. *N Engl J Med.* 2003;348(17):1639-1646.
16. Sandhu R, Finkelhor RS, Gunawardena DR, Bahler RC. Prevalence and characteristics of left ventricular noncompaction in a community hospital cohort of patients with systolic dysfunction. *Echocardiography.* 2008;25(1):8-12.
17. Zaragoza MV, Arbustini E, Narula J. Noncompaction of the left ventricle: primary cardiomyopathy with an elusive genetic etiology. *Curr Opin Pediatr.* 2007;19(6):619-627.
18. Ichida F, Tsubata S, Bowles KR, et al. Novel gene mutations in patients with left ventricular noncompaction or Barth syndrome. *Circulation.* 2001;103(9):1256-1263.
19. Bione S, D'Adamo P, Maestrini E, Gedeon AK, Bolhuis PA, Toniolo D. A novel X-linked gene, G4.5. is responsible for Barth syndrome. *Nat Genet.* 1996;12(4):385-389.
20. Sasse-Klaassen S, Gerull B, Oechslin E, Jenni R, Thierfelder L. Isolated noncompaction of the left ventricular myocardium in the adult is an autosomal dominant disorder in the majority of patients. *Am J Med Genet A.* 2003;119(2):162-167.
21. Bartram U, Bauer J, Schranz D. Primary noncompaction of the ventricular myocardium from the morphogenetic standpoint. *Pediatr Cardiol.* 2007;28(5):325-332.
22. Sedmera D, Pexieder T, Vuillemin M, Thompson RP, Anderson RH. Developmental patterning of the myocardium. *Anat Rec.* 2000;258(4):319-337.

23. Wessels A, Sedmera D. Developmental anatomy of the heart: a tale of mice and man. *Physiol Genomics.* 2003;15:165-176.
24. Aras D, Tufekcioglu O, Ergun K, et al. Clinical features of isolated ventricular noncompaction in adults long-term clinical course, echocardiographic properties, and predictors of left ventricular failure. *J Card Fail.* 2006;12(9):726-733.
25. Agmon Y, Connolly HM, Olson LJ, Khandheria BK, Seward JB. Noncompaction of the ventricular myocardium. *J Am Soc Echocardiogr.* 1999;12(10):859-863.
26. Jenni R, Wyss CA, Oechslin EN, Kaufmann PA. Isolated ventricular noncompaction is associated with coronary microcirculatory dysfunction. *J Am Coll Cardiol.* 2002;39(3):450-454.
27. Buonanno C, Variola A, Dander B, Gabaldo S, Marafioti V. Isolated noncompaction of the myocardium: an exceedingly rare cardiomyopathy. A case report. *Ital Heart J.* 2000;1(4):301-305.
28. Weiford BC, Subbarao VD, Mulhern KM. Noncompaction of the ventricular myocardium. *Circulation.* 2004;109(24):2965-2971.
29. Stollberger C, Finsterer J. Left ventricular hypertrabeculation/noncompaction and stroke or embolism. *Cardiology.* 2005;103(2):68-72.
30. Stollberger C. Finsterer J. Thrombi in left ventricular hypertrabeculation/noncompaction – review of the literature. *Acta Cardiol.* 2004;59(3):341-344.
31. Frischknecht BS, Attenhofer Jost CH, Oechslin EN, et al. Validation of noncompaction criteria in dilated cardiomyopathy, and valvular and hypertensive heart disease. *J Am Soc Echocardiogr.* 2005;18(8):865-872.
32. Hany TF, Jenni R, Debatin JF. MR appearance of isolated noncompaction of the left ventricle. *J Magn Reson Imaging.* 1997;7(2):437-438.
33. Petersen SE, Selvanayagam JB, Wiesmann F, et al. Left ventricular noncompaction: insights from cardiovascular magnetic resonance imaging. *J Am Coll Cardiol.* 2005;46(1):101-105.
34. Daimon Y, Watanabe S, Takeda S, Hijikata Y, Komuro I. Two-layered appearance of noncompaction of the ventricular myocardium on magnetic resonance imaging. *Circ J.* 2002;66(6):619-621.
35. Soler R, Rodríguez E, Monserrat L, Alvarez N. MRI of subendocardial perfusion deficits in isolated left ventricular noncompaction. *J Comput Assist Tomogr.* 2002;26(3):373-375.
36. Alsaileek AA, Syed I, Seward JB, Julsrud P. Myocardial fibrosis of left ventricle: Magnetic resonance imaging in noncompaction. *J Magn Reson Imaging.* 2008;27(3):621-624.
37. Jenni R, Rojas J, Oechslin E. Isolated noncompaction of the myocardium. *N Engl J Med.* 1999;340(12):966-967.

Abstract of Interest

Clinical features of isolated ventricular noncompaction in adults long-term clinical course, echocardiographic properties, and predictors of left ventricular failure

ARAS D, TUFEKCIOGLU O, ERGUN K, ET AL.
ANKARA, TURKEY

BACKGROUND: Isolated ventricular non-compaction (IVNC) is a rare disorder characterized by prominent trabecular meshwork and deep recesses. We retrospectively assessed the clinical characteristics and natural course of IVNC in adults diagnosed at our hospital.

METHODS AND RESULTS: Sixty-seven adult patients (44 male, mean age 41 ± 18 years) with the diagnosis of IVNC were evaluated in this retrospective cohort. Its prevalence was found to be 14%. Forty-seven patients (70%) had class I/II functional capacity. Fifty-seven patients (85%) had electrocardiographic abnormalities, and the most common one was left ventricular (LV) hypertrophy (25%). LV systolic function was depressed in 44 patients (66%), with a median ejection fraction (EF) of 35% (range: 20%-48%) at diagnosis. Multiple regression analysis revealed that age at initial presentation, the total number of affected segments, and the ratio of non-compaction/compaction (NC/C) were the independent predictors of LV systolic dysfunction. Familial occurrence of IVNC was 33%. During a mean follow-up of 30 months (range: 9-50 months), major complications including ventricular tachycardia, heart failure requiring hospitalization, and cerebrovascular events were observed in 36%, 34%, and 9% of the patients, respectively. Ten patients (15%) with IVNC died in this study. LVEF at initial presentation and functional capacity at last visit were found to be independent predictors of mortality.

CONCLUSION: This study suggests that IVNC is a form of cardiomyopathy with higher prevalence and relatively better prognosis than previously reported. Age at initial presentation, ratio of NC/C, and number of affected segments seem to be major determinants of LV systolic dysfunction, while initial LVEF and last functional capacity predict mortality in this cohort.

J Card Fail. 2006;12(9):726-733.

Upcoming Meetings

30 April – 2 May 2008
Quality of Care and Outcomes Research in Cardiovascular Disease and Stroke Conference 2008
Baltimore, Maryland
Contact: http://www.americanheart.org/presenter.jhtml?identifier=3049494
E-mail: scientificconferences@heart.org
Telephone: (888) 242-2453 or (214) 570-5935

1 – 3 May 2008
European Society of Cardiology (ESC) EuroPRevent 2008
Paris, France
Contact: ww.escardio.org/

30 August – 3 September 2008
ESC Congress 2008
Munich, Germany
Contact: www.escardio.org/

Disclosure Statement: *Drs. Thavendiranathan and Chow have stated that they have no disclosures to announce in association with the contents of this issue.*

Change of address notices and requests for subscriptions to *Cardiology Rounds* are to be sent by mail to P.O. Box 310, Station H, Montreal, Quebec H3G 2K8 or by fax to (514) 932-5114 or by e-mail to info@snellmedical.com. Please reference *Cardiology Rounds* in your correspondence. Undeliverable copies are to be sent to the address above. Publications Post #40032303