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## CASE REPORT

### Biventricular Hypertrophic Cardiomyopathy in a 26-year-old Nigerian Woman with Noonan Syndrome

*Cardiomyopathie Hypertrophique Biventriculaire Chez une Nigériane de 26 Ans Atteinte du Syndrome de Noonan.*

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#### ABSTRACT

**BACKGROUND:** Cardiac disorders are found in about half of cases of Noonan syndrome (NS). The most common congenital heart diseases in this syndrome include pulmonary valvular stenosis (60%), interatrial septal defect (25%), and obstructive or non-obstructive hypertrophic cardiomyopathy (17%). Biventricular hypertrophic cardiomyopathy (HCM) is very rare in this condition.

**OBJECTIVE:** The objective is to report a case of biventricular hypertrophic cardiomyopathy in a 26-year-old Nigerian female with the phenotype.

**METHODS:** This is a descriptive case report.

**RESULTS:** The patient presented with dyspnoea on exertion which started at the age of 7 years and has progressively worsened. There was associated precordial chest pain and palpitation.

Clinical examination revealed a young woman, who is small for her age. She had some dysmorphic features such as a webbed neck, low-set ears, low posterior hairline, crowded teeth, high arched palate, a small and asymmetric chin and a high carrying angle at the elbows.

The pulses were synchronous and there was no radio-radial or radio-femoral delay and her blood pressures were within normal limits. Cardiac auscultation was unremarkable.

The 12-lead ECG showed biventricular hypertrophy with a strain pattern. The echocardiogram showed features in keeping with biventricular hypertrophic cardiomyopathy.

**CONCLUSION:** Biventricular HCM is relatively uncommon in Noonan syndrome. Patients with typical dysmorphia should have a full cardiac evaluation to look for these anomalies.

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**KEYWORDS:** Noonan syndrome, Dysmorphia, Hypertrophic cardiomyopathy.

#### RÉSUMÉ

**CONTEXTE:** Des troubles cardiaques sont présents chez environ la moitié des patients atteints du syndrome de Noonan. Les cardiopathies congénitales les plus fréquentes dans ce syndrome incluent la sténose valvulaire pulmonaire (60 %), la communication interauriculaire (25 %) et la cardiomyopathie hypertrophique obstructive ou non obstructive (17 %). La cardiomyopathie hypertrophique (HCM) biventriculaire est très rare dans ces conditions.

**OBJECTIF:** L'objectif est de rapporter un cas de cardiomyopathie hypertrophique biventriculaire chez une femme nigériane de 26 ans présentant le phénotype.

**MÉTHODES:** Il s'agit d'un rapport de cas descriptif.

**RÉSULTATS:** La patiente s'est présentée avec une dyspnée à l'effort, débutant à l'âge de 7 ans et s'aggravant progressivement. Il y avait des douleurs précordiales associées ainsi que des palpitations.

L'examen clinique a révélé une jeune femme, petite pour son âge, avec quelques traits dysmorphiques tels qu'un cou palmé, des oreilles basses, une implantation basse des cheveux à l'arrière, des dents serrées, un palais ogival, un menton petit et asymétrique et un angle de port élevé au niveau des coudes.

Les pouls étaient synchrones, sans retard radio-radial ou radio-fémoral, et la tension artérielle était dans les limites normales. L'auscultation cardiaque n'a révélé aucune anomalie.

L'électrocardiogramme à 12 dérivations a montré une hypertrophie biventriculaire avec un pattern de strain. L'échocardiogramme a révélé des caractéristiques compatibles avec une cardiomyopathie hypertrophique biventriculaire.

**CONCLUSION:** La cardiomyopathie hypertrophique biventriculaire est relativement rare dans le syndrome de Noonan. Les patients présentant une dysmorphie typique devraient subir une évaluation cardiaque complète afin de détecter ces anomalies.

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**MOTS CLÉS:** Syndrome de Noonan, Dysmorphie, Cardiomyopathie hypertrophique.

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