

MOGE(S) nosology in low-to-middle-income countries

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In a very thoughtful News and Views article (Mayosi, B. M. MOGE(S): a standardized classification of cardiomyopathies? *Nat. Rev. Cardiol.* **11**, 134–135 [2014]),¹ Professor Mayosi described MOGE(S) as an “admirable achievement on the path towards a globally accepted nomenclature for cardiomyopathy”.² He appropriately emphasized the lack of examples of aetiological notation for important cardiomyopathies from low-income and middle-income countries, such as tropical endomyocardial fibrosis (EMF; also known as tropical endomyocardial disease, constrictive endocarditis, and endocarditis parietalis fibroplastica), which is one of the most prevalent causes of restrictive cardiomyopathy.³ Mayosi suggests that an

aetiological category for unexplained (or idiopathic) cardiomyopathy would improve the clinical utility of the MOGE(s) scheme.¹ The authors of the scheme completely agree, and reiterate that the classification system (or nosology) is flexible, conveniently expandable, and allows easy modification of the MOGE(S) online app.⁴ The first update to the app has been already proposed to improve the description of arrhythmogenic right ventricular cardiomyopathy.⁵

EMF might provide a paradigmatic example of the flexibility of the MOGE(S) nosology and associated app. EMF is estimated to be the underlying cause of 20% of heart failure in the areas of Africa in which EMF is endemic, and is known to be the second leading cause of hospital admissions

for acquired cardiovascular disease in children and young adults, after rheumatic heart disease.⁶ However, the aetiology of EMF remains elusive. Several hypotheses have been proposed, such as cardiotoxicity of eosinophils and hypereosinophilia (†Eo), often associated with parasitic infectious aetiology (schistosomiasis, *Loa loa* microfilaria, filariasis, or malaria), autoimmunity (high titres of malarial antibodies, hyperimmune malarial splenomegaly, raised anti-streptolysin titres in patients with EMF), genetic or ethnic predisposition (high incidence among particular ethnic groups in some countries), diet (tuber cassava, tapioca) in association with extreme deprivation of proteins (tryptophan deficiency), and geochemical and climate characteristics (caesium derived from monazite soils).^{7–14} Although none of these factors has been proven to cause EMF, the MOGE(S) nosology can be invoked as follows.

The condition can manifest as isolated or dominant left ventricular (LV) EMF, isolated or dominant right ventricular (RV) EMF, or biventricular (RV + LV) EMF, and can be stated as such in the MOGE(S) description. MOGE(S) can also describe the organ involvement in addition to the heart (H), such as liver involvement (H + L), or †Eo. Aetiology can be listed as being known (K), suspected (S), or unknown (U). If known or suspected, the aetiology can be coupled with description of the injurious agent. Although isolated EMF is easily described by MOGE(S), the presence of †Eo goes beyond the cardiac evaluation and requires haematological and immunological investigation and classification. Various classifications of †Eo disorders and associated syndromes have been formulated in the past, and the Year 2011 Working Conference on Eosinophil Disorders and Syndromes refined the criteria and definitions.¹⁵ The description of EMF with †Eo, with or without involvement of organs other than the heart (such as lungs, skin, liver, gastrointestinal tract, pancreas, or kidneys)

Box 1 | MOGE(S) classifications of tropical EMF

M_{R (EMF LV)} **O**_{H+†Eo} **G**_N **E**_{I-S-Schistosomiasis} **S**_{C-IV}

M Restrictive cardiomyopathy (R) associated with EMF predominantly involving the left ventricle (LV)
O Heart involvement (H) and †Eo
G Absence of familial aggregation (N)
E Infectious aetiology (I), possibly schistosomiasis, which is suspected (S) as the cause of the EMF
S AHA stage C (structural heart disease with previous or current symptoms of heart failure), in NYHA functional class IV

M_{R (EMF RV+LV)} **O**_{H+Li+†Eo} **G**_N **E**_{I-LU} **S**_{C-III}

M Restrictive cardiomyopathy (R) associated with EMF involving both the right ventricle (RV) and the left ventricle (LV)
O Heart (H) and liver (Li) involvement and †Eo
G Absence of familial aggregation (N)
E Possible infectious aetiology (I), which is actually unknown (U)
S AHA stage C, in NYHA functional class III

M_{R (EMF RV)} **O**_{H+Li+†Eo} **G**_N **E**_{I-S-Helminthiasis} **S**_{C-IV}

M Restrictive cardiomyopathy (R) associated with EMF predominantly involving the right ventricle (RV)
O Heart (H) and liver (Li) involvement and †Eo
G Absence of familial aggregation (N)
E Infectious aetiology (I), possibly helminthiasis, which is suspected (S) as the cause of the EMF
S AHA stage C, in NYHA functional class IV

M_{R (EMF LV)} **O**_{H+†Eo} **G**_N **E**_{Eo-HES-PDGFRB/PDGFRB} **S**_{C-II}*

M Restrictive cardiomyopathy (R) associated with EMF predominantly involving the left ventricle (LV)
O Heart (H) involvement and †Eo
G Absence of familial aggregation (N)
E Eosinophilic heart disease (Eo) and hypereosinophilic syndrome (HES), in which a *PDGFRA/PDGFRB* fusion gene has been identified
S AHA stage C, in NYHA functional class II

*This example is provided to describe the EMF variety associated with hypereosinophilic syndrome and mutations in oncogenic tyrosine kinase receptors. Abbreviations: EMF, endomyocardial fibrosis; †Eo, hypereosinophilia.

requires a multidisciplinary diagnostic work-up, starting with elucidation of the cause of †Eo. The important information for treatment is the identification of fusion genes involving *PDGFRA*, *PDGFRB*, and *FGFR1*. MOGE(S) can describe the mutations in oncogenic tyrosine kinase receptors in the aetiology section, or their absence (negative), or the lack of information (not tested).

The examples in Box 1 were generated using the MOGE(S) app.⁴ Therefore, MOGE(S) and its app can readily provide a description for any type of cardiomyopathy, with the advantage of integrating important information that can constitute the basis for a novel, worldwide epidemiology to describe diseases, not only on the basis of their clinical phenotypes, but also according to their aetiology.

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Competing interests

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