

## CORRELATIONS BETWEEN SYSTEMIC FEATURES AND DENTAL ABNORMALITIES IN MITRAL VALVE PROLAPSE SYNDROME

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### ABSTRACT

**Aim of the study** To establish if dental anomalies are linked to systemic features for patients with mitral valve prolapse syndrome (MVPS). **Material and methods** We've compared the dental findings from 27 patients diagnosed with MVPS with the dental features from 27 patients without MVPS. Complete dental, cardiological and psychological consults were performed. Biochemical and haematological laboratory findings were followed. Electrocardiogram, echocardiogram, dental X-ray confirmed clinical diagnosis. Treatment received by patients with MVPS consisted in betablockers, dental procedures and psychological interventions. **Results** MVPS was associated with a higher prevalence of dental anomalies and negative impact among life quality. **Conclusions** Future researches could reveal algorithms for better understanding of MVPS and dental associated anomalies, with clinical and practical consequences.

**Keywords:** mitral valve prolapsed syndrome, dental anomalies, systemic features

### INTRODUCTION

Mitral valve prolapse was first described by John Barlow, in 1966, as Barlow's syndrome (1). It's a clinical condition due to myxomatous degeneration of the mitral valve, sometimes followed by hemodynamical consequences: severe mitral regurgitation.

Last decade studies confirmed the fact that MVPS is a genetic disorder, include in so called fibrillinopathies (2). The defect is due to mutations or deletions of gene encoding fibrillin 1 (located on chromosome 15). An

abnormal fibrillin could affect stretch and elasticity of connective tissue from heart valves, aorta, eyes, skin, joints. There are different phenotype expressions of the disease: Marfan syndrome, MASS (mitral prolapsed valve, borderline aorta dilation, skin and skeletal features) phenotype, mitral valve prolapse syndrome and only skeletal marfanoid habitus features (3).

Oral manifestations in fibrillinopathies are represented by disorders of temporo-mandibular joints, maxillary protrusion, high

palate, crowded teeth(4). Enamel hypoplasia is another modification of dental structure and there is a higher prevalence of periodontal inflammation in these patients (5). Oral anomalies are due to abnormal fibrillin 1, which composes extracellular microfibrils, in association with elastin or in elastin-free bundles(6). Risk of infective endocarditis related to oral pathology and associated valvulopathies is more prominent at these patients. Practical consequences are related to a good oral care, concerning infective endocarditis prophylaxis.

## MATERIAL AND METHODS

Our study included 56 patients, divided in 2 groups: group A, composed by 27 patients with MVPS and group B, composed by 27 patients without MVPS. The study developed during 12 months. We've selected control group (group B) to have the same gender distribution as group A (80% females) and similar ages( between 7-38 years old).

All patients were clinical consulted by a cardiologist, a dentist and a psychologist. The cardiologist also performed electrocardiogram and transthoracic echocardiography; skeletal modifications were followed according to Ghent criteria (7). MVPS has been defined if the patient had both mitral valve prolapse (echocardiography) and skeletal features of marfanoid habitus. Echocardiography was useful for hemodynamical consequences of mitral valve prolapse assessment: severity of mitral regurgitation, systolic and diastolic left ventricle functions, associated pulmonary arterial hypertension- acceleration time of systolic pulmonary flow, systolic pulmonary artery pressure. Electrocardiography followed associated tachyarrhythmias and discrete changes due to left ventricular performances.

Betablockers were recommended if tachyarrhythmias were electrocardiographical documented and after the female patients

were advised to avoid pregnancy during this treatment.

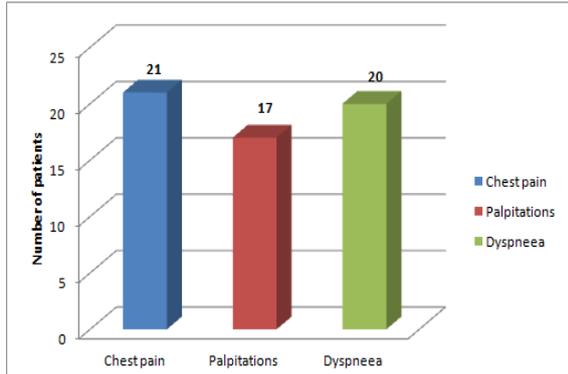
Biological and haematological findings were about liver function (transaminases), kidney function (serum creatinine), electrolyte balance (serum calcium, natrium, potassium, magnesium), glycaemia, complete blood count.

Dental consultation followed International Headache Society's criteria for temporomandibular joint (TMJ) disorder (7), enamel defects, periodontal inflammation, teeth abnormalities. Only 1 patient required X-ray investigation for temporomandibular joint, for the others clinical criteria were sufficient in diagnosing dysfunction of this joint. Specific dental treatment (orthodontic, endodontic) and physical therapy addressed to temporomandibular joint dysfunction were performed through dental office. Oral rehabilitation was very important, because it provided a self-respect increasing, a possibility for adequate alimentation, esthetical improvement and prophylaxis against infective endocarditis.

Psychological consultation used conversation and HAMA (Hamilton anxiety) scale for psychiatric status assessment. Cognitive Behaviour Therapy was applied to selected patients suitable for this treatment.

## RESULTS AND DISCUSSIONS

Group A, with MVPS revealed a higher incidence of dental anomalies than group B, without MVPS. We also followed to select adequate treatment for cardiological and dental disorders. Among all 27 patients of group A, the cardiovascular symptoms were represented by atypical chest pain (80% from 27-21 patients), palpitations (65% from 27-17 patients) and dyspnea: 76% from 27-20 patients (fig.1).



**Figure 1. Cardiovascular symptoms in group A (MVPS)**

Skeletal features for marfanoid habitus were revealed in all 27 patients from group A; wrist and/or thumb sign : 20% from 27 - 5 patients; pectus excavatum : 25% from 27- 6 patients; pectus carinatum : 14% from 27- 3 patients; thoraco-lumbar skolyosis and /or cyphosis : 38% from 27- 10 patients; facial features (retrognathia, dolicocephalia, malar hypoplasia, enophthalmia, downslating palpebral fissures) : 32% from 27- 8 patients.

Echocardiography confirmed mitral valve prolapse in all 27 patients from group A and quantified severity mitral regurgitation .We discovered 57% from 27 - 15 patients with mild valvulopathy; 29% from 27- 7 patients had moderate mitral regurgitation (fig. 2 and fig. 3) and only 14% from 27- 2 patients had severe valvulopathy.



**Figure 2. Echocardiography: posterior and anterior mitral valve prolapse**



**Figure 3. Echocardiography: moderate mitral regurgitation**

These 2 last patients had echocardiographical modifications due to secondary pulmonary arterial hypertension (acceleration time pulmonary systolic flow-78 msec and 65 msec, respectively - normal values being higher than 100 msec; systolic pulmonary artery pressure- 56 mmHg and 48 mmHg, respectively- normal values between 25-35 mmHg). They refused surgical valve replacement recommendations.

Electrocardiogram revealed ventricular premature beats in 37% from 27- 9 patients and an atrial fibrillation episode in 1 patient with severe mitral regurgitation. All these 10 patients were up to 16 years and they received betablockers (Bisoprolol 5mg/day), being advised to avoid pregnancy during this treatment.

About laboratory findings there were 2 patients with mild anemia due to prolonged menstruations and they received substitution treatment with Ferrous sulphate 100mg /day for 6 months. They were advised to consult an endocrinologist and a gynecologist.

Dental consultation discovered temporomandibular joint dysfunction in 35% from group A - 9 patients and only in 5% from group B - 1 patient. These patients had the following criteria (8): recurrent pain in one or more region of the head/ face; evidence that pain is due to temporomandibular joint dys-

function: precipitating factors of pain –mandible movements /chewing, diminished mandible opening, click sound from temporomandibular joint during mandible movements, tenderness of this joint; headache resolution after dysfunction joint treatment.

The questions and physical examination for temporomandibular joint disorder were performed at the beginning of the study and after that, every 3 months; the dentist noticed if there were ameliorations in TMJ function.

Enamel defects were presented in 46% from group A - 12 patients and only 23% from group B - 6 patients. Teeth abnormalities (vicious implantation) were revealed in 25% from group A - 6 patients (fig. 4 and fig. 5) and in 5% from group B - 1 patient.



Figure 4. Dental anomalies: malocclusion



Figure 5. Dental anomalies: abnormal implantation

Periodontal inflammation had a higher prevalence among group A - 58% from 27 - 15 patients, comparing with group B - 7% from 27 - 2 patients. The differences between group A and B concerning dental anomalies are expressed in figures 6 and 7.

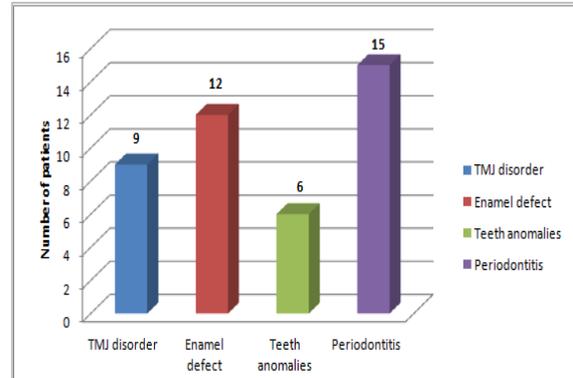


Figure 6. Dental anomalies in group A (MVPS)

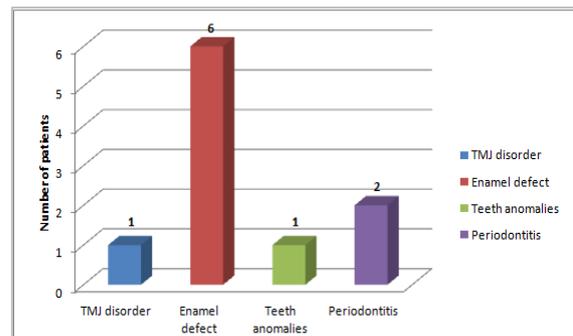


Figure 7. Dental anomalies in group B (without MVPS)

All these patients received specific dental treatment and they all improved their oral and general health.

Psychological distress was more prominent in group A than in group B ( a 26 average HAMA scale for group A comparing to a 12 average HAMA scale for group B: mild anxiety - higher than 18; moderate anxiety- higher than 25; severe anxiety- higher than 30).There were 80% from group A - 21 patients with psychological interventions and favourable evolution.

## CONCLUSIONS

1. Patients with MVPS had a higher prevalence of dental pathology than patients without MPVS; this observation is con-

cordant to other studies from medical literature according Marfan syndrome (9,10,11).

2. The originality of our study is due to observation of the patients with a type of fibrillinopathy - MVPS, different as a phenotype from Marfan syndrome.

3. Future researches are necessary for

establishing a more powerful link between cardiovascular and dental abnormalities in these genetic disorders.

4. Multidisciplinary collaboration could be an important objective in our future projects, as a continuance of this present work.

## REFERENCES

- 1 Barlow JB, Bosman CK, Aneurysmal protrusion of the posterior leaflet of the mitral valve. An auscultatory-electrocardiographic syndrome. *Am Heart J.* 1966; 71 (2): 166-78
- 2 Nasuti JF, Zhang PJ, Feldman MD, Fibrillin and other matrix proteins in mitral valve prolapse syndrome. *Ann Thorac Surg.* 2004 ;77 (2): 532-6.
- 3 Biggin A1, Holman K, Brett M, Detection of thirty novel FBN1 mutations in patients with Marfan syndrome or a related fibrillinopathy. *Hum Mutat.* 2004 ; 23(1): 99.
- 4 De Coster PJA, Martens LCM, De Paepe A, Oral manifestations of patients with Marfan syndrome: A case-control study. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2002; 93(5): 564-572
- 5 Staufenbiel I, Hauschild C, Kahl-Nieke B, Periodontal conditions in patients with Marfan syndrome—a multicenter case control study. *BMC Oral Health.* 2013, 13:59
- 6 Sugawara Y, Sawada T, Inoue SJ, Immunohistochemical localization of elastin, fibrillins and microfibril-associated glycoprotein-1 in the developing periodontal ligament of the rat molar. *Periodontal Res.* 2010 ; 45(1): 52-9
- 7 Loeys BL, Dietz HC, Braverman AC, The revised Ghent nosology for the Marfan syndrome. *J Med Genet.* 2010, 47: 476-485.
- 8 Bauss O, Sadat-Khonsari R, Fenske C, Temporomandibular joint dysfunction in Marfan syndrome. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2004, 97: 592-598.
- 9 Ammash NM, Sundt TM, Connolly HM, Marfan syndrome-diagnosis and management. *Curr Probl Cardiol.* 2008, 33: 7-39.
- 10 Westling L, Mohlin B, Bresin A, Craniofacial manifestations in the Marfan syndrome: palatal dimensions and a comparative cephalometric analysis. *J Craniofac Genet Dev Biol* 1998, 18: 211-218.
- 11 Rybczynski M, Bernhardt AM, Rehder U, The spectrum of syndromes and manifestations in individuals screened for suspected Marfan syndrome. *Am J Med Genet A.* 2008, 146A:3157-3166.