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Duplication of Interventricular Septum and a Double Orifice Mitral Valve resulting in a three-ventricular chamber heart: A most rare anomaly

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Introduction (or Basis or Objectives):
The human heart derives from embryonic mesodermal germ-layer cells that differentiate into mesothelium, endothelium, and myocardium and starts with formation of two endocardial tubes, which merge, loop and septate to form the four chambers. It starts to beat by the 4th week, at the end of which two atrioventricular endocardial cushions appear to form the atrio-ventricular valves. An intra-muscular septum then grows within the ventricle mass consisting of a membranous and a muscular part.

Methods &Results:
We assessed a 63-year-old woman with a history of intermittent chest pains, palpitations and shortness of breath on moderate exertion. She had a complex previous history of mild systemic lupus combined with Sjogrens Syndrome, skin angiokeratomas and osteomalacia with Vitamin D deficiency. She also had a dual chamber pacemaker implanted for 2:1 heart block. Initially an echocardiogram demonstrated a thickened interventricular septum with a pattern that followed asymmetric septal hypertrophy and normal left ventricular (LV) systolic function. The septal wall showed marked hypertrabeculation and subsequent cardiovascular magnetic resonance (CMR) imaging demonstrated bulky almost exophytic-looking trabeculae at the septal wall and the global LV mass was 1.4 times normal. Moreover both echocardiography and CMR imaging revealed that the thickened interventricular septum was divided into two muscle bands from the apex to the base giving the appearance of septal duplication and of a three-ventricular chamber heart (Figures 1). Interestingly, duplication of the atrio-ventricular valves was also noted with an appearance of double orifice mitral valve. The most septal valve showed dysplastic leaflets and restricted opening into the rudimentary septal chamber.

Conclusions:
We describe this unusual case of congenital heart disease where the first impression was suggestive of hypertrophic cardiomyopathy or cardiomyopathy with asymmetric septal hypertrophy and non-compaction appearance. CMR with delayed gadolinium enhancement imaging did not show enhancement compatible with metabolic or infiltrative disorders. Myocardial biopsy unfortunately is not available as the patient did not consent to undergo the procedure. Duplication of the interventricular septum and formation of a third rudimentary ventricular cavity is extremely rare congenital abnormality. To our knowledge this has never been described before.