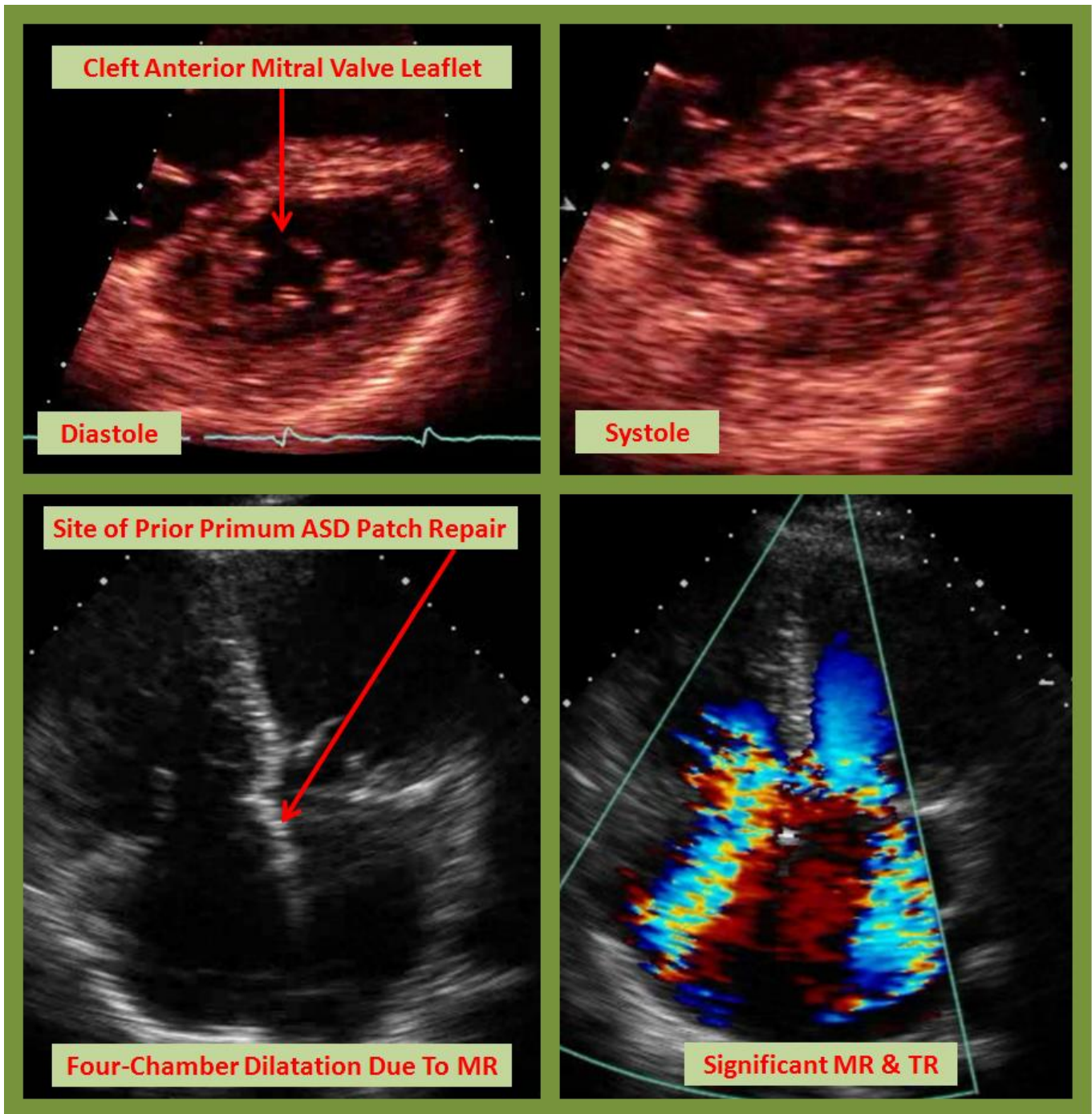


Cleft Mitral! A Deleterious Primum Residue!

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Description

Atrial Septal Defect (ASD) is the most common congenital cardiac anomaly in the adult. The most common form is the ostium secundum, which is usually an isolated congenital anomaly, and often discovered incidentally. Sinus venosus ASD is associated with anomalous connection of the right upper pulmonary vein, while primum ASD is frequently associated with a cleft anterior mitral valve leaflet causing significant mitral regurgitation (MR). It is crucial to be aware of such associations in planning surgery, so that repair of these lesions is performed at the time of ASD closure [1].

Primum ASD (partial atrioventricular canal defect) can be accompanied by a variety of anomalies of the endocardial cushion, most prominently a cleft mitral and/or tricuspid valve leaflet. It can also be associated with left-sided obstructive lesions such as coarctation, subaortic stenosis, a single papillary muscle (parachute mitral valve causing congenital mitral stenosis) or double orifice left ventricle, and can present with significant CHF symptoms if not detected and repaired early. These findings may be seen in up to 10% of patients with primum ASD [2].

Short- and long-term survival for repair of isolated primum ASD approaches 100% with need for reintervention, typically directed at management of MR, in only 5 to 10%. Complete repair involving closure of the primum atrial septal defect and repair of the cleft of the mitral valve with pericardial patch has been the practice since 1982 [3].

Patients, who do not undergo closure of the cleft, can have significant mitral valve regurgitation later in life. The postoperative grade of MR is an independent risk factor for late significant MR, so even mild regurgitation postoperatively should be addressed. While late development of moderate-to-severe MR can either be repaired or replaced, the outcomes are better with early repair. A challenging technical characteristic for the surgeon is the ability to maintain the mitral valve competency in the postoperative period [4].

Therefore, a high index of suspicion should be maintained in the work-up and planning for surgery in atrial septal defects, so that associated lesions can be detected with the use of echocardiography, CT angiography and cardiac MRI, in addition to identifying a skilled surgeon capable of performing such complex repairs [5].

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