Introduction: An atrial appendage to ventricular accessory pathways are uncommon. It can be congenital or acquired (surgically created). The epicardial approach may be required for the successful ablation of such a pathway due to the epicardial course. We report a successful ablation of a rare case of ‘epicardial accessory pathway’ (AP) connecting the right atrial appendage (RAA) to the right ventricular outflow tract (RVOT).

Methods: A 19-year-old boy with dysmorphic facies, cleft palate, pectus excavatum, mental retardation and a structurally normal heart presented with recurrent palpitations and a history of failed RFA. The 12-lead sinus rhythm ECG was suggestive of ventricular pre-excitation. An orthodromic AVRT was reproducibly inducible. Maximal pre-excitation by atrial pacing suggested right free wall anteriorly located AP.

Result: Extensive mapping, with the aid of fluoroscopy and 3-D electro-anatomic imaging, along the tricuspid annulus during maximal preexcitation and AVRT did not yield satisfactory A-V potentials. Mapping was also performed in the coronary aortic cusps. Mapping within the right atrial appendage (RAA) yielded the most satisfactory signals including sharp AP potentials. The RAA anatomy was diligently defined using angiogram and 3-D mapping. RF lesions (40 W; 43ocelcius; 8 seconds) during sinus rhythm resulted in the separation of A-V potentials and loss of pre-excitation. Overdrive atrial and ventricular pacing demonstrated absence of AP. There was no inducible tachycardia. At 6-month follow-up, there was no recurrence. The location of the successful site of ablation is indicative of congenital epicardial A-V connection, atrial appendage being the atrial end and adjoining right ventricular outflow tract as the ventricular end.

Conclusion: One should suspect atrial appendage to ventricular connections if preexcitation shows QS pattern in V1 & V2 and inferior axis in limb leads especially in a case of previously attempted RF ablation.