This study details the finding of a novel missense variant in filamin A gene (*FLNA)* identified through whole-exome sequencing in a case of familial Ebstein’s anomaly (EA)*.*These results provide evidence for a highly penetrant identifiable genetic cause for this condition in the six living affected blood relatives diagnosed with EA. Though rare, familial EA has been reported previously, including in association with mutations in *MYH7* and *NKX2.5*, both of which are inherited in an autosomal dominant way*.  FLNA* is located on the X chromosome and the X-linked inheritance of this novel variant is consistent with the phenotypes observed in this family. An X-linked pattern of inheritance for EA was not previously reported in the literature. Appreciation of the different inheritance patterns of familial EA is important in order to give accurate counseling regarding the recurrence risks for this condition.  In addition, the clinical details in this report expand the phenotypic spectrum associated with mutations in *FLNA*.   The recognition of these additional features will allow the clinician to conduct comprehensive clinical evaluation when assessing individuals with pathogenic variants in this gene.  Such features may also act as diagnostic clues when assessing those with EA and a possible underlying genetic cause.