

Title: *FBN1*-Related Marfan Syndrome *GeneReview* Table 1  
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 Date: February 2022

**Table 1. Calculation of the Systemic Score**

Feature	Value	Enter Value if Feature Is Present
Wrist AND thumb sign	3	
Wrist OR thumb sign	1	
Pectus carinatum deformity	2	
Pectus excavatum or chest asymmetry	1	
Hindfoot deformity	2	
Plain flat foot (pes planus)	1	
Pneumothorax	2	
Dural ectasia	2	
Protrusio acetabulae	2	
Reduced upper segment / lower segment AND increased arm span/height ratios	1	
Scoliosis or thoracolumbar kyphosis	1	
Reduced elbow extension	1	
3 of 5 facial features	1	
Skin striae	1	
Myopia	1	
Mitral valve prolapse	1	
<b>Total</b>		

A systemic score calculator and a complete description of each component evaluation can be found at the [Marfan Foundation](#) website.