Prevalence of marfan syndrome from 2006 to 2013in the Korean

Marfan syndrome is a genetic disorder of connective tissue. Marfan syndrome is caused by mutations in the *FBN1* gene on chromosome 15, which encodes fibrillin-1, a glycoprotein component of the extracellular matrix. Fibrillin-1 is essential for the proper formation of the extracellular matrix, including the biogenesis and maintenance of elastic fibers. The extracellular matrix is critical for both the structural integrity of connective tissue, but also serves as a reservoir for growth factors Elastic fibers are found throughout the body.Clinical presentation for Marfan syndrome ranges from mild to severe systemic disease. If marfan syndrome patients are not correctly managed, it may lead to early or sudden death. It's because Marfan syndrome involves in the circulatory system such as mitral valve prolapse, aortic regurgitation, a dilated aorta, an aortic aneurysm, or an aortic dissection.

The objective of the study is to show prevalence of marfan syndrome between 2006 and 2013 in Korean.

Data were collected from Korean National Health Insurance Benefit records from 2006 through 2013.

The age-standardized prevalence of congenital heart diseases in adults was calculated with the direct method using the estimated Korean population in 2010 as the reference.

Overall, the age-standardized cumulative prevalence of marfan syndrome was 0.92 per 100,000 persons in 2006 and 2.32 in 2013. For female, the age-standardized cumulative prevalence of marfan syndrome was 0.70 per 100,000 persons in 2006 and 1.92 in 2013. For male, the age-standardized cumulative prevalence of marfan syndrome was 1.08 per 100,000 persons in 2006 and 2.61 in 2013.

The overall age-standardized cumulative prevalence of marfan syndrome increased from about 1 to about 3 persons per 100,000 persons between 2006 and 2013. The teens showed higher prevalence than that of other age groups, especially in 15-19 years males and 10-14 years females. Males showed slightly higher prevalence of marfan syndrome than that of female.