CONCLUSION FH is a grave ailment with its genesis in early childhood resulting in damaging consequences in later life.The past 2 decades have noted the evolution of novel therapies to lower LDL–cholesterol levels and defer premature atherosclerosis, especially in conjunction with lifestyle modifications.While such treatment initiatives have notably improved the prognosis of HeFH, the outcomes of familial homozygous hypercholesterolemia remain disappointing.Despite these triumphs, a large majority of children do not attain targeted lipid goals due to shortfalls in diagnosis, monitoring, and treatment.Early initiation of lipid–lowering therapy and lifestyle measures might improve the clinical outcome.Although most cases may be treated with a combination of statins and cholesterol absorption inhibitors, some will have need of more invasive therapies such as LDL apheresis.An effective screening strategy together with timely initiation of established therapies would go a long way in reducing the burden of atherosclerosis due to this challenging condition.Although the need for a screening strategy to detect this disease early is widely accepted, there is no consensus regarding whom and when to screen.