

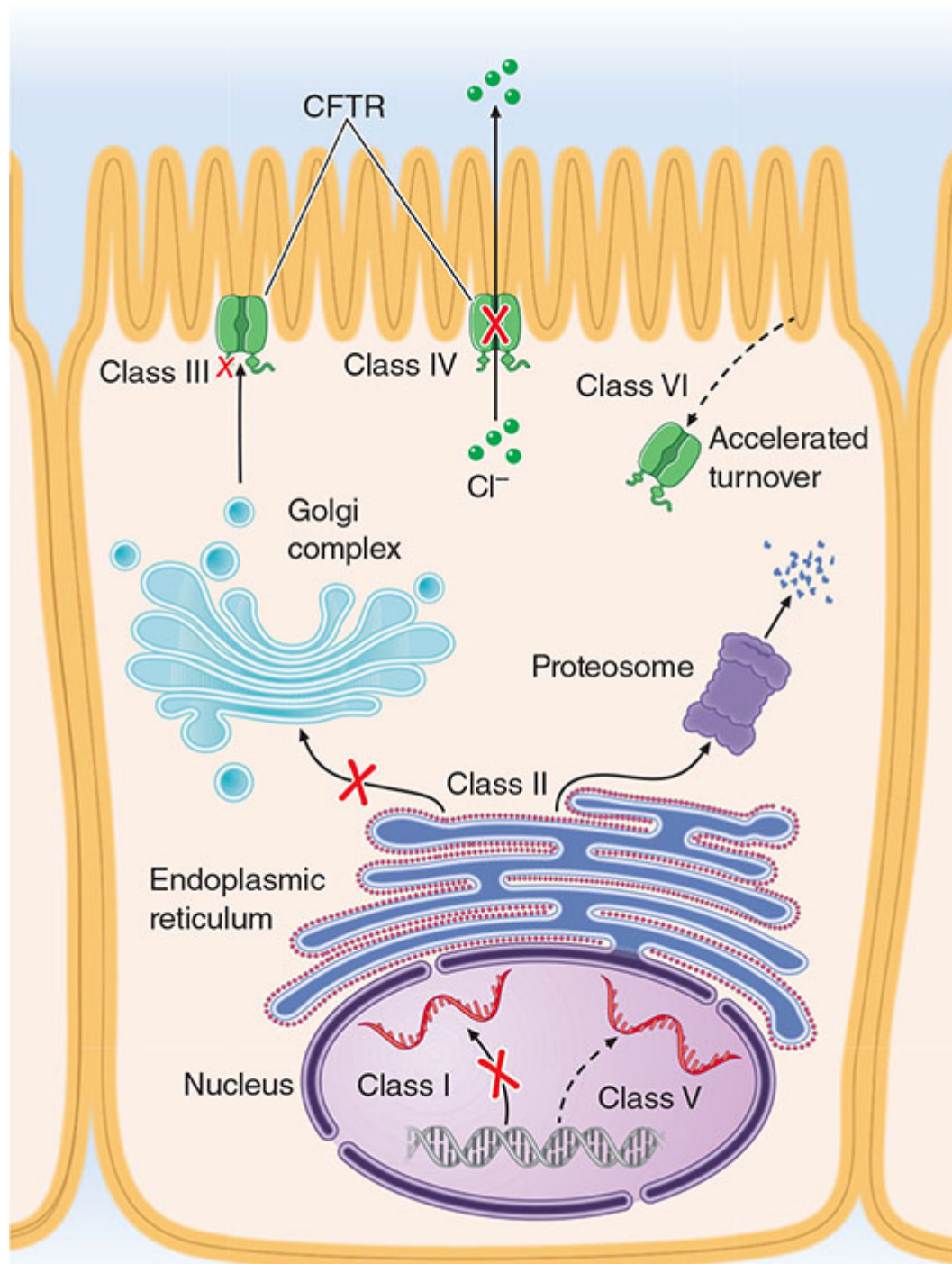


FIGURE 291-2 Categories of *CFTR* mutations. Classes of defects in the *CFTR* gene include the absence of synthesis (class I); defective protein maturation and premature degradation (class II); disordered gating/regulation, such as diminished adenosine triphosphate (ATP) binding and hydrolysis (class III); defective conductance through the ion channel pore (class IV); a reduced number of *CFTR* transcripts due to a promoter or splicing abnormality (class V); and accelerated