

Self Administration of PERT Medication in students with Cystic Fibrosis

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Introduction

Cystic Fibrosis (CF) is the most common inherited, autosomal recessive disorder in Caucasians. Approximately 1 in 2500 live births result in CF which causes an underlying abnormality of the chloride ion channel. This abnormality impacts most organ systems, with the G.I and respiratory systems most severely impacted. While CF is still considered a terminal disorder, life expectancy has increased dramatically!

This favorable finding has resulted in many more children with CF living into adolescence and young adulthood. The management of adolescents with CF therefore poses new and specific challenges for school nurses in the middle and high school setting.

Recognizing how CF negatively impacts an adolescent's biological, mental and psychosocial health issues is essential knowledge for school nurses in order to promote the successful management of CF in this population.

Objectives

Identify how CF may impact body image, risk taking behavior, independence and nonadherence to therapy regimes in the adolescent student.

Discuss challenges students with CF often encounter due to symptoms associated with CF.

Determine how self carry legislation of PERT medication may help adolescents with CF develop more independent functioning.

Cystic Fibrosis and its Impact

Many CF health issues become more “visible” during adolescent years producing embarrassing symptoms such as coughing, flatulence, steatorrhea, short stature, low body mass and pubertal delay.

Challenges

The symptoms associated with CF negatively impact treatment adherence and body image. Symptoms also create barriers to achieving independence and feelings of self worth

Body Image: 60% of adolescents with CF are distressed by visible body/maturation differences
Depression/Anxiety: Symptoms are 2 -3 times greater than community sampled peers.

Nonadherence to treatment: 50% of adolescents with CF have periods of nonadherence to treatment and taking medication.

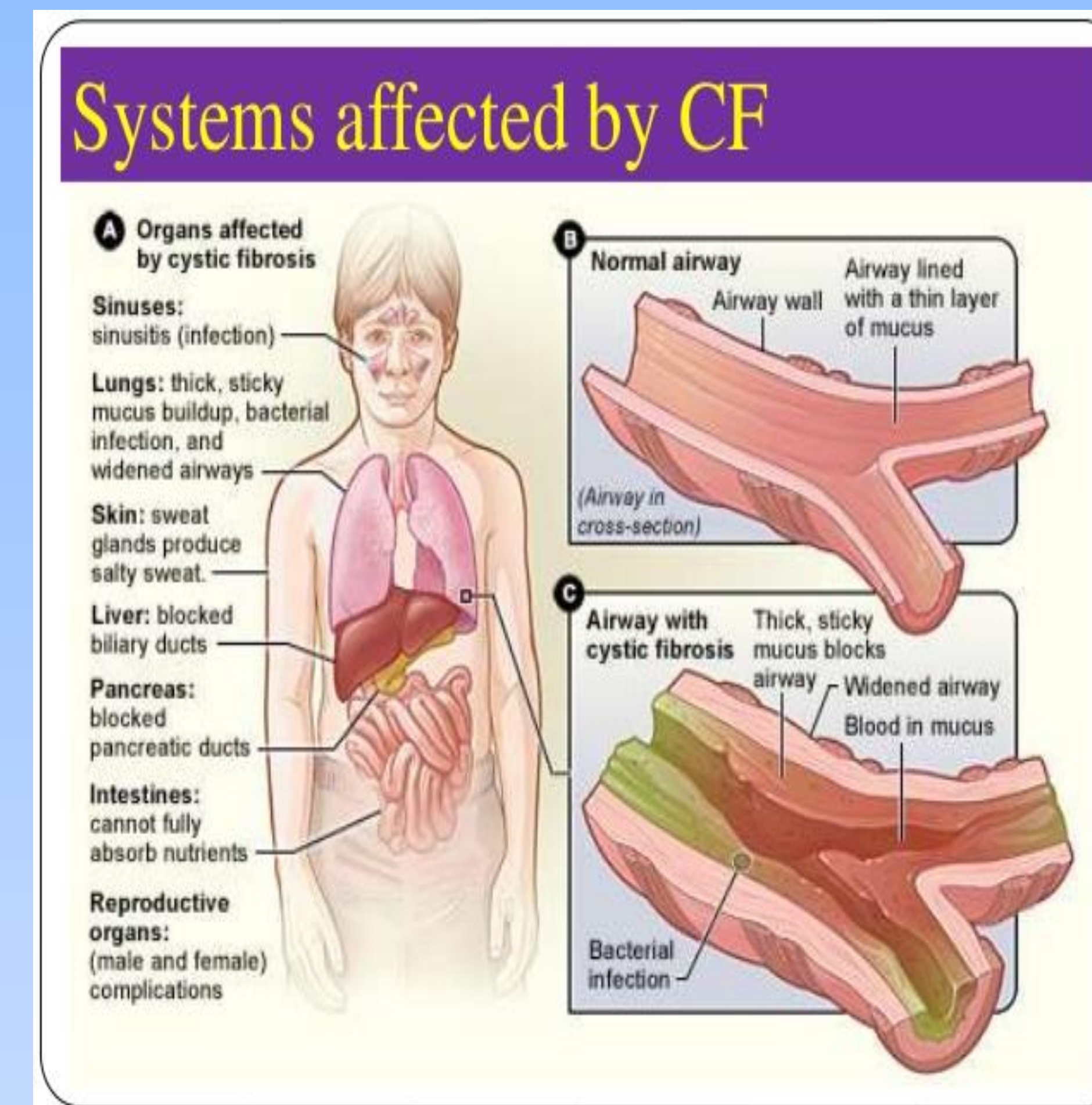
Fertility: Males (98%) have azoospermia (leads to decreased use of contraception). Females sub-fertile (increases risky behavior).

Risk Taking Behaviors: Elevated compared to same aged peers. Preventing illness and reducing risk of infection is key role for school nurse

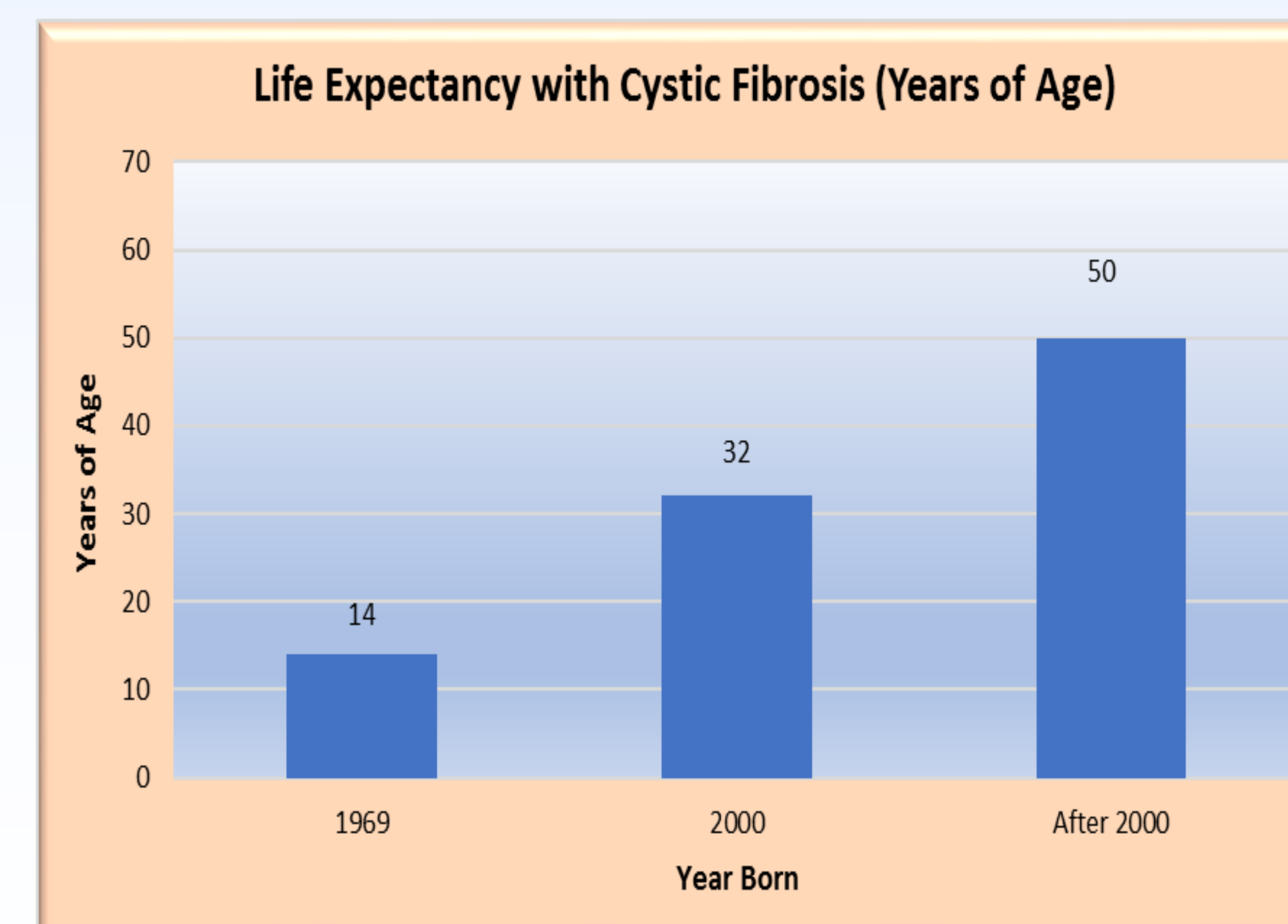
Independence: Altered due to treatment burden, enzyme therapy regime, infection protection protocols.

Uncertainty of Life Expectancy: Lower self worth than peers and higher rate of mental illness.

Disease Impact



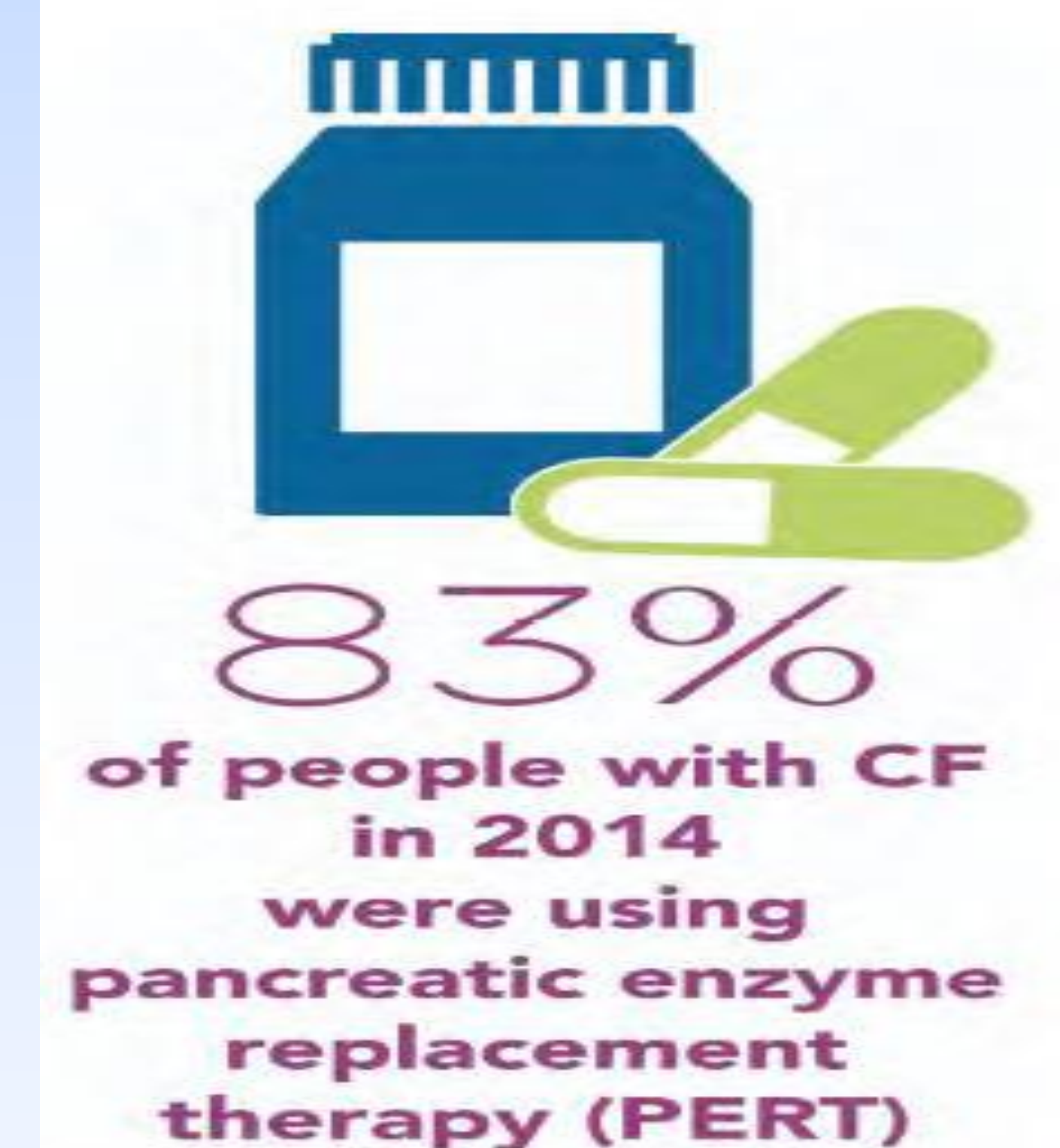
The literature examining health outcomes in adolescents with Cystic Fibrosis indicates that students with CF feel different. New Legislation permitting students to self carry their PERT medication lessens the treatment burden, promotes adherence to medication therapy and increases independent functioning in this population.



Conclusions

The literature examining health outcomes in adolescents with Cystic Fibrosis indicates that these individuals often feel self-conscious, isolated and powerless, making this already tumultuous time of life more challenging. School Nurses are positioned to implement strategies that counter these barriers to typical development. Promoting independence through self administration of PERT medications is one such strategy Illinois school nurses can now use.

Self Carry Laws



References

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