

# Pseudocholinesterase deficiency

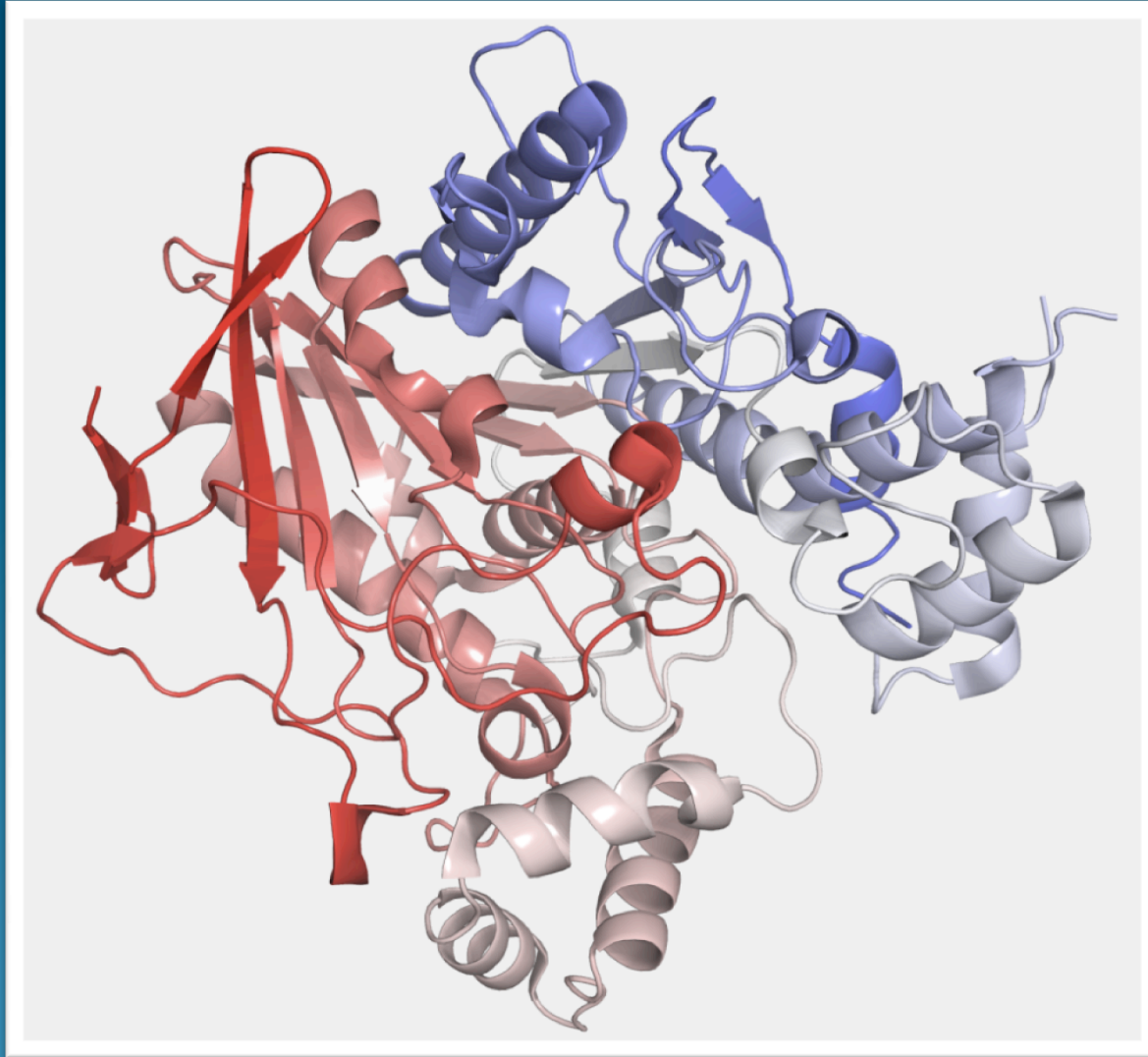
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# What is Pseudocholinesterase?

- Also called Plasma Cholinesterase
- Enzyme in Plasma
- Succinylcholine is metabolized in bloodstream
- Normal Enzyme metabolizes Sux
- Not found in NM Junction
- Synthesized in liver
- Severe liver dysfunction, i.e. > 25% is needed to decrease enzyme level to abnormal levels

# Model of Molecule





# Effects on Other Drugs

- Also involved in metabolism of
  - Ester Local Anesthetics
  - Mivacurium



# Succinylcholine Apnea

- Relaxation can last 3-6 hours after dosing
- Abnormal Quality of Enzyme
- Very slow metabolism of Sux
- Prolonged relaxation after Sux administration
- Prolongation of effect of sux can also be caused by decreased concentrations of pseudocholinesterase, causing prolonged blockade
- Echothiophate, Organophosphates can also permanently block the enzyme's activity
- In absence of enzyme function, apnea resolves by slow renal elimination



# Genetics

- Chromosome 3, single locus
- 4 Alleles:
  - Usual; (Eu)
  - Atypical (Ea)
  - Silent (Absent); (Es) Silent gene incapable of metabolizing enzyme
  - Fluoride Resistant (Ef)
- 10 genotypes

Genotype	Incidence	Duration of Block	Dibucaine Number	Fluoride Number
Eu:Eu	96%	Normal	80	60
Eu:Ea	1:25	+	60	45
Eu:Es	1:90	+	80	60
Eu:Ef	1:200	+	75	50
Ea:Ea	1:2800	++++	20	20
Ea:Ef	1:20000	++	50	35
Es:Ea	1:29000	++++	20	35
Es:Es	1:100000	++++	-	-
Ef:Es	1:150000	++	60	35
Ef:Ef	1:154000	++	70	30



# Dibucaine Number

- Local Anesthetic used to test enzyme activity
- Dibucaine inhibits normal enzyme
- Eu:Eu (normal) form inhibited by 80%
- Ea:Ea inhibited by 20%
- Heterozygous forms vary in Dibucaine Number-see chart
- Usual part of evaluation and work-up





# Fluoride Number

- Fluoride inhibits normal enzyme
- “Fluoride Number” also available as lab test for evaluation and work-up



# Evaluation

- Family History
- Lab Work would include:
  - Dibucaine Number
  - Fluoride Number
  - Pseudocholinesterase Concentration
- Note- these tests require being sent to a reference lab, and turn-around time is several days



# Treatment

- If susceptible patient receives succinylcholine, the only treatment is ventilatory support until the neuromuscular blockade is gone.




# Question

You are in the holding room talking to your first patient of the day. He was not seen in pre-op clinic. He is a 90 kg 35 y.o. man who is having an open inguinal herniorrhaphy. He is a farmer, smokes 1 ppd, and takes atenolol for mild HTN. The remainder of his personal history and physical exam, including his airway exam, are normal (MP II, normal dentition and ROM). He did have a tonsillectomy when he was 6 years old without any apparent problems.

When you asked him about any family history of anesthetic-related problems, he informs you that he thinks his brother was allergic to some anesthetic drug. This episode happened 10 years ago, His brother lives out of state now.

He is curious that you are concerned about this information, and he really wants this procedure done today, as this is his slow season on the farm.

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- What are your concerns?
- What would be in your differential diagnosis?
- What other questions would you ask the patient?
- Is there any further workup that you can do, or is needed?
- Do you have any options to proceed with this patient's anesthetic?

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- What are your concerns?
  - What would be in your differential diagnosis?
  - What other questions would you ask the patient?

The patient says he can call his mother and ask her for more details. You hand him a phone. What questions do you ask his mother?

- Is there any further workup that you can do, or is needed?
- Do you have any options to proceed with this patient's anesthetic?
- How do you counsel patient?



# References

- Longnecker DE, et al: Principles and Practices of Anesthesiology. McGraw-Hill Professional, 2007
- Peck TE, et al: Pharmacology for anaesthesia and intensive care. Cambridge University Press, 2003
- Miller RD, et al: Miller's Anesthesia, Edition: 6; Elsevier Churchill Livingstone, 2005