

The pyruvate dehydrogenase complex (PDC) catalyzes the conversion of pyruvate to acetyl-CoA, a substrate for the Krebs (citric acid) cycle. The rate of pyruvate conversion is greatly reduced in individuals with PDC deficiency, a rare disorder.

(a) **Identify** the cellular location where PDC is most active.

Identification (1 point)

- Mitochondria
- Mitochondrial matrix

a. PDC is most active in a cell's mitochonoria.



FRQ Friday #8

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(b) Make a claim about how PDC deficiency affects the amount of NADH produced by glycolysis AND the amount of NADH produced by the Krebs (citric acid) cycle in a cell. Provide reasoning to support your claims based on the position of the PDC-catalyzed reaction in the sequence of the cellular respiration pathway.

(1 point per row; 2 points max.)

	Claim	Reasoning	
Glycolysis	No change	Glycolysis continues; PDC is not needed.	
		Glycolysis occurs before conversion of pyruvate to acetyl-CoA.	
Krebs cycle	Decrease	 The Krebs cycle is greatly reduced/slowed down if there is no/less acetyl-CoA. 	
		The Krebs cycle occurs after conversion of pyruvate to acetyl-CoA.	



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	Claim	Reasoning	
Glycolysis	No change	Glycolysis continues; PDC is not needed.	
		 Glycolysis occurs before conversion of pyruvate to acetyl-CoA. 	
Krebs cycle	Decrease	The Krebs cycle is greatly reduced/slowed down if there is no/less acetyl-CoA.	
		The Krebs cycle occurs after conversion of pyruvate to acetyl-CoA.	

b. A PDC deficiency woes not change the amount of
NADH produced by glycolysis, but it decreases
the amount of NADH produced in the Krebs cycle.
to make acetyl con
This occurs because the PDC-cortalyzed reaction
occurs after glyrolysis, leading to roimpact, and
before the Krebs cycle. Without acetyl COA, the Krebs
cycle cannot occur, so a PDC deficiency would halt
all NADH production in this stop.



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(c) PDC deficiency is caused by mutations in the *PDHA1* gene, which is located on the X chromosome. A male with PDC deficiency and a homozygous female with no family history of PDC deficiency have a male offspring. **Calculate** the probability that the male offspring will have PDC deficiency.

	Х	X
X	XX	XX
Y	XY	XY

Calculation (1 point)

- The probability of inheritance is 0.
- The offspring cannot/will not have PDC deficiency.



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c. Male - Xmy, where m= mutation					
Female - XX	There is a 0% probability				
<u> </u>	that a male offspring :				
	will have PDC deficiency.				

