

Newsletter of Science Society May, 2014

二零一四年五月號

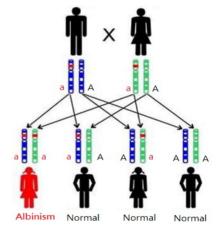
+++++ Albinism +++++

白化病(albinism)是一種較常見的皮膚及其附屬器官黑色素 (Melanin) 缺乏所引起的疾病,由於先天性缺乏酪氨酸酶,或酪氨酸酶功能減退,黑色素合成發生障礙所導致的遺傳性白斑病。這類病人通常是全身皮膚、毛髮、眼睛缺乏黑色素,因此表現為眼睛視網膜 (Retina) 無色素,虹膜 (Iris) 和瞳孔 (Pupil) 呈現淡粉色,怕光,看東西時總是眯著眼睛。皮膚、眉毛、頭髮及其他體毛都呈白色或白裡帶黃。

白化病的發病是由於黑色素代謝障礙所致。正常人體內的黑色素由黑色素細胞合成,黑色素細胞內有黑素小體,它含有酪氨酸酶 (Tyrosinase) ,這種酶能將酪氨酸轉變成黑色素。白化病患者體內黑色素細胞數目正常,細胞內也有黑素小體,但由於控制酪氨酸酶的基因發生突變,不能合成酪氨酸酶,於是黑素小體中酪氨酸酶缺乏,不能使酪氨酸轉變成黑色素,從而導致皮膚、粘膜、毛髮、眼等白化。

白化病屬於家族遺傳性疾病,為常染色體隱性遺傳 (Autosomal recessive disorder) ,常發生於近親結婚的人群中。白化病遺傳圖譜:患者雙親均攜帶白化病基因(Aa),本身不發病。如果夫婦雙方同時將所攜帶的致病基因(aa)傳給子女,子女就會患病。而子女中男女患病機會均等,子女的發病幾率是 1/4。

白化病可分為兩大群,一為較常見的眼皮膚白化病,機體不能製造 黑色素。另一類為伴有異常免疫系統的白化病,與黑色素及其他細胞蛋白 的缺陷有關。



1.眼白化病(Ocular Albinism, OA)

病人僅眼色素減少或缺乏,具有不同程度的視力低下,畏光等症狀。眼白化病為X連鎖隱性遺傳, 是由母親所攜帶的白化病基因傳給兒子時才患病,傳給女兒一般不患病。

2.眼皮膚白化病(Oculocutaneous Albinism, OCA)

除眼色素缺乏和視力低下、畏光等症狀外,病人皮膚和毛髮均有明顯色素缺乏。

3.白化病相關綜合徵

病人除具有一定程度的眼皮膚白化病表現外,還有其他異常,如同時具有免疫功能低下的 Chediak-Higashi綜合徵和具有出血素質的Hermansky-Pudlak綜合徵,這類疾病較為罕見。

白化病全身皮膚缺乏黑色素而呈乳白或粉紅色,柔嫩發乾。 毛髮變為淡白或淡黃。由於缺乏黑色素的保護,患者皮膚對光線高 度敏感,日晒後易發生曬斑和各種光感性皮炎而皮膚曬後不變黑。 也常發生光照性唇炎、毛細血管擴張,有的發生日光性角化,並可 發生基底細胞癌或鱗狀細胞癌。眼部由於色素缺乏,虹膜為粉紅或 淡藍色,常有畏光、流淚、眼球震顫及散光等症状。大多數白化病 患者體力及智力發育較差。



Screaming for Ice Cream

Summer, where I'm from, is a wonderful thing. When the weather warms up, people head outdoors. Days are long and hot—perfect conditions for canoeing, biking, and having picnics by the lake. Best of all, a sweaty brow is a great excuse to gather your friends and go out for a drippy cone of ice cream.

It's cold. It's sweet. It's creamy. And that burst of fruit-filled, nutty, or chocolate-chunky flavor can be incredibly refreshing when the steamy heat of midday starts to weigh you down. As far as I'm concerned, ice cream is summer's most delightful treat.

Not all ice cream, however, is perfect ice cream. Texture matters as much as flavor.

Nothing's worse than an icy scoop, or one that tastes grainy, syrupy, or artificial.



So, what's the secret to decadent ice cream that tastes like a dream and feels like a silky cloud melting in your mouth? High-quality ingredients, for one, are essential, plus fine-tuned techniques(微調技術) that combine ingredients in just the right proportions with a perfect amount of air whipped in.

"There's a lot of science behind it," says David Smith. He's a food scientist at the University of Minnesota, Twin Cities.

For ice-cream manufacturers, the science of ice cream matters a lot. According to the International Ice Cream Association, people in the United States spend more than \$20 billion on cold, creamy treats each year. The U.S. Department of Agriculture says that, in 2004, each person ate an average of about 21.5 quarts of ice cream.

To satisfy the population's unceasing demand for the sweet stuff, companies are constantly on the lookout for ways to make better-tasting ice cream that lasts longer, costs less, and is more nutritious than current varieties.

Topnotch ingredients

Besides cream, ice cream has just a few essential ingredients: mainly sugar, milk solids, ice crystals, air, and flavorings. Sugar makes the dessert sweet, but it also serves another important purpose. In the freezer, plain cream turns into a solid that's hard as a rock. Sugar lowers the mixture's freezing temperature, making it much softer.

The highest quality ice creams have the fewest ingredients. From vanilla extract to fresh strawberries, each component is topnotch(最高級的).



The best ice cream varieties also tend to have the least air in them, which makes them denser. A cheap brand may be half air, Smith says. Gourmet brands are more like 15 to 20 percent air. In other words, the better the ice cream, the more of it you actually get in each bite.

"Pick up a half gallon of economy brand and a quart of super-premium," Smith says. Even though the half-gallon is much bigger when it comes to volume, there's not much difference in the amount of the frozen stuff that you get. Air is pumped into ice cream near the end of the manufacturing process, after the basic ingredients have been mixed together and cooled down, but before fillings, chunks, and other flavorings go in.

As the concoction freezes in a huge container, large blades spin the creamy goo around and scrape ice crystals off the sides of the container. For high-end brands with lots of butterfat (乳脂), the process is enough to prevent iciness. Some companies churn their ice cream slowly and for a long time. This process helps fat globules stick together and produces a creamy, somewhat greasy texture.

Economy brands that skimp on richness and are churned more quickly, however, have to add extra ingredients. Emulsifiers(乳化劑), for example, keep fat suspended (懸浮) throughout the final product. And stabilizers(穩定劑) control the growth of ice crystals.

Some companies don't use stabilizers. Left in the freezer for too long, a carton of this sort of ice cream ends up with an icy beard on top. Ice cream that melts and refreezes often has the same problem.

Yummy and healthy

In the battle against ice crystals, one recent avenue of research has focused on molecules called antifreeze (抗凝劑) proteins. Found in certain types of fish and plants that live in extremely cold environments, these proteins prevent ice crystals from forming, which keeps the organisms from freezing to death. Perhaps they could do the same for ice cream. The technique isn't yet practical, however.

In the meantime, many companies are trying hard to make ice cream that is both yummy and healthy. In its traditional form, ice cream is loaded with calories and fat. It's the fat that carries the flavor and produces the smooth texture. "You've got to have fat," Smith says.

All that fat, however, is a problem when it comes to obesity, diabetes, heart disease, and other weight-related illnesses. So far, no one has managed to create a low-fat version of ice



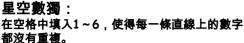
cream that tastes as good as the real thing. That's because normal ice cream is about 60 to 62 percent water and 10 to 20 percent butterfat, Smith says. (Ten percent is the minimum amount of butterfat a product must contain to qualify as true ice cream.)

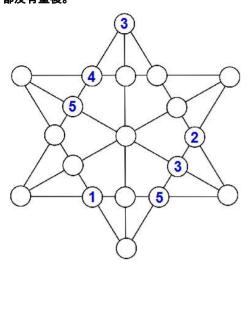
Once you start taking out fats, water content can shoot up to 70 to 78 percent, Smith says. The more water a frozen dessert contains, the quicker it turns to ice and slush, and the less flavor it has.

While it's not a good idea to have sundaes and milkshakes with every meal, a cool cone may be one of the most enjoyable parts of summer, and it's not necessary to give up the habit altogether. When you do indulge, just make sure to savor every bite. Appreciating it fully might make the experience even more special.

"Ice cream is unique because you don't get it every day," Smith says. "You eat it on special occasions or when you go to the park. It's usually a happy time."

~~ TIME TO RELAX ~~



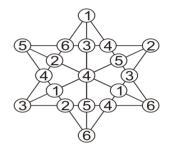


<u>Sodoku</u>

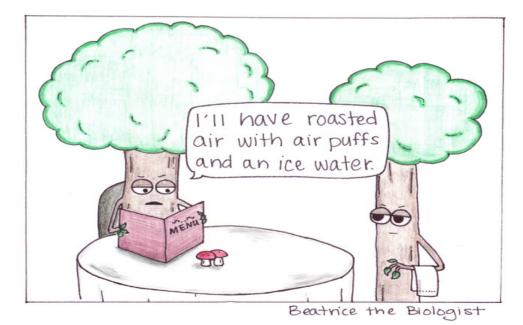
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	7					4		2
4				5		9	3	
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	1	6		8				4
8		1					4 7	
	3	5		1	6		7	
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Previous Sodoku Answer

9	2	3	6	7	4	5	8	1
5	8	7	2	9	1	6	3	4
6	4	1	8	5	3	9	7	2
8	3	2	5	6	9	1	4	7
1	9	6	7	4	8	3	2	5
7	5	4	1	3	2	8	9	6
2	6	8	9	1	7	4	5	3
4	7	5	3	8	6	2	1	9
3	1	9	4	2	5	7	6	8



Comic Corner: Tree Restaurant



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