

## 红细胞6-磷酸葡萄糖脱氢酶 缺乏者的白细胞活酶

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红细胞6-磷酸葡萄糖脱氢酶(G 6 P D)缺乏与蚕豆病、药物溶血及新生儿高胆红素血症发病有密切关系。广东省是G 6 P D缺乏的高发地区之一,广东籍侨民G 6 P D缺乏者白细胞酶活性的检查已有报告<sup>(1,2)</sup>,但国内尚未见类似研究。本文报告19例G 6 P D缺乏者白细胞酶活性的检查结果。

### 材 料 和 方 法

19例G 6 P D缺乏者,是采用红细胞G 6 P D活性测定法检出,均为成年人。除1例为广西籍人外,余18例为广东人。红细胞G 6 P D活性测定是用杜氏的改良法<sup>(3)</sup>。白细胞酶制备根据Bonsignore<sup>(4)</sup>的方法稍作改良。为避免酶损失,白细胞分离后不作低渗清除污染红细胞的处理。白细胞悬液置-30℃反复冻结和融冻4次,0℃10,000×g离心15分钟,上清液用于酶活性测定,方法与红细胞酶活性测定相同。同时测定白细胞溶解液中污染的血红蛋白量,并据红细胞G 6 P D活性测定的结果,从白细胞酶活性测定结果中扣除与白细胞溶解液中污染的血红蛋白量相当的红细胞酶活性。G 6 P D缺乏以红细胞G 6 P D活性低于正常对照平均值的45%(2.71单位)作为判断标准。19例中有2例女性红细胞G 6 P D活性在正常范例围(18、19),是根据家系研究确认为酶缺乏的杂合子。

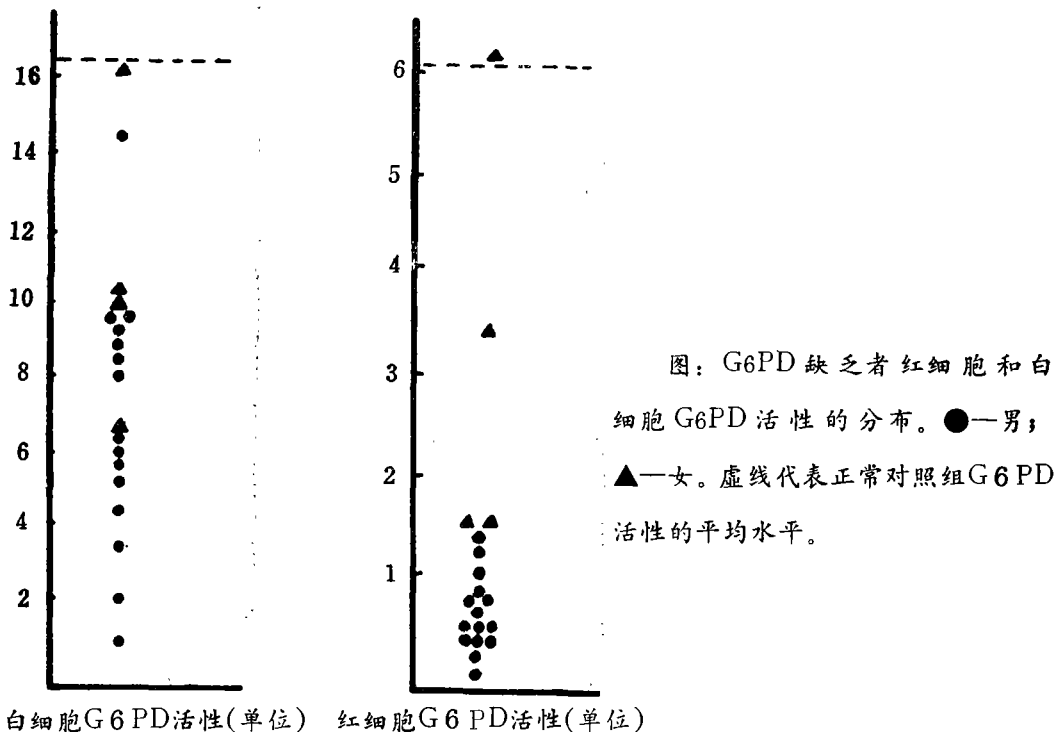
### 结 果

G 6 P D缺乏者红细胞和白细胞G 6 P D活性及其分布,见表和图。

表 G 6 PD 缺乏者的红细胞和白细胞 G 6 PD 活性

| 病 例 号           | 性 别 | 红细胞 G 6 PD 活性(单位)                                    | 白细胞 G 6 PD 活性(单位)                                       |
|-----------------|-----|------------------------------------------------------|---------------------------------------------------------|
| 1               | 男   | 0.18                                                 | 1.05                                                    |
| 2               | "   | 0.34                                                 | 6.20                                                    |
| 3               | "   | 0.54                                                 | 9.11                                                    |
| 4               | "   | 0.55                                                 | 6.60                                                    |
| 5               | "   | 0.75                                                 | 14.90                                                   |
| 6               | "   | 0.75                                                 | 8.04                                                    |
| 7               | "   | 0.79                                                 | 2.19                                                    |
| 8               | "   | 0.79                                                 | 9.64                                                    |
| 9               | "   | 0.79                                                 | 4.67                                                    |
| 10              | "   | 0.88                                                 | 8.83                                                    |
| 11              | "   | 0.98                                                 | 3.54                                                    |
| 12              | "   | 1.00                                                 | 5.79                                                    |
| 13              | "   | 1.16                                                 | 5.85                                                    |
| 14              | "   | 1.36                                                 | 9.86                                                    |
| 15              | "   | 1.51                                                 | 9.88                                                    |
| 16              | 女   | 1.73                                                 | 16.52                                                   |
| 17              | "   | 1.77                                                 | 6.72                                                    |
| 18              | "   | 3.58                                                 | 10.11                                                   |
| 19              | "   | 6.26                                                 | 10.79                                                   |
| 平均              |     | $1.35 \pm 1.4$                                       | $7.91 \pm 3.8$                                          |
| 非 G 6 PD 缺乏正常对照 |     | $6.02 \pm 1.4$ (67例)<br>(2.92~8.19;<br>男性 2.92~8.19) | $16.48 \pm 5.6$ (58例)<br>(6.16~32.80;<br>男性 8.99~32.80) |

从图中可见, G 6 PD 缺乏者红细胞和白细胞 G 6 PD 活性的分布较红细胞 G 6 PD 活性的分布分散。在 15 例男性 G 6 PD 缺乏者, 红细胞 G 6 PD 活性为  $0.82 \pm 0.35$  单位, 所有 15 例均低于正常对照组平均值的 25%, 属酶严重缺乏类型。15 例白细胞 G 6 PD 活性为  $7.08 \pm 3.5$  单位, 患者酶活性虽均在正常平均水平以下, 但有相当数量的患者酶的活性在正常对照组正常值的下限之上。在 4 例女性 G 6 PD 缺乏者, 经家系研究检出的 2 例属酶缺乏的杂合子, 红细胞酶活性在正常范围, 另 2 例红细胞酶活性亦相当于酶缺乏的杂合子水平。4 例红细胞 G 6 PD 活性平均为  $3.34 \pm 2.1$  单位, 白细胞 G 6 PD 活性平均为  $11.04 \pm 4.0$  单位。就个体而言, 不论是红细胞或白细胞 G 6 PD 活性均可达到正常平均水平。



19例 G6PD 缺乏者白细胞 G6PD 活性显著低于 58 例正常对照 ( $P < 0.001$ ), 为正常对照组酶活性的 48%, 与文献所报告中国人 G6PD 缺乏者白细胞 G6PD 活性为正常对照均值的 49%<sup>(1)</sup> 和 44%<sup>(2)</sup> 相近似。

15 例男性或全部 19 例 G6PD 缺乏者, 其红细胞 G6PD 活性与白细胞 G6PD 活性相关均无显著性 (15 例男性  $r = 0.347$ ,  $P > 0.1$ ; 19 例  $r = 0.326$ ,  $P > 0.5$ )。

## 讨 论

在有关 G6PD 缺乏与蚕豆病、药物溶血及新生儿高胆红素血症的关系的研究中, G6PD 缺乏个体一般都是通过红细胞酶活性检查而检出, 少有同时测定有核细胞如白细胞 G6PD 活性者。本文报告有红细胞 G6PD 活性严重缺乏者, 白细胞酶活性虽亦有显著缺乏, 但个体酶活性分布范围广泛, 相当部分个体酶活性在正常对照组正常值的下限之上。而红细胞 G6PD 和白细胞 G6PD 活性无平行关系, 红细胞酶活性缺乏程度不反映白细胞酶活性水平, 原因之一可能是该组 G6PD 缺乏者实际上包含了多种不同的遗传变异型。已知不同的 G6PD 缺乏变异型对蚕豆病、药物溶血及新生儿高胆红素血症有不同的易感性。如黑人中最常见的 G6PD 缺乏变异型 G6PDA- 罕见发生蚕豆病, 其白细胞和肝细胞 G6PD 活性则仅有轻度降低。广东地区 G6PD 缺乏者食用蚕豆后亦只部分人发病。白细胞与其他有核细胞如肝细胞 G6PD 活性的关系, 肝 G6PD 活性与肝解毒功

能及其与蚕豆病、药物溶血、新生儿高胆红素血症发病的关系仍未见研究、值得探讨。

### 参 考 文 献

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## Leukocyte Enzyme Activity in Erythrocyte Glucose - 6 - Phosphate Dehydrogenase Deficiency

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### Abstract

Leukocyte G 6 PD activity was studied in 19 subjects with erythrocyte G 6 PD deficiency. All are natives of Guangdong Province except one. In 15 males G 6 PD activities in erythrocytes averaged  $0.82 \pm 0.35$  IU and the enzyme activities of all 15 cases were 25% lower than the average value of normal controls ( $6.02 \pm 1.4$  IU). Leukocyte G 6 PD activity was  $7.08 \pm 3.5$  IU and below the average of normal ( $16.48 \pm 5.6$  IU). However, in a number of individuals the enzyme activities of leukocytes were above the inferior limit of normal controls. In females, 2 of 4 were heterozygotes of G 6 PD deficiency identified by family studies, G 6 PD activities of erythrocytes were found to be within the normal range. In the other 2 cases, the erythrocyte enzyme activities fell into the enzyme level of G 6 PD deficient heterozygotes. The average G 6 PD activity of 4 females was  $3.34 \pm 2.1$  IU

for erythrocytes and  $11.04 \pm 4.0$  IU for leukocytes.

The average G 6 PD activity in leukocytes of 19 subjects was significantly lower, about 48% of the normal value, than that of normal controls, and this result was similar to that reported by others previously.

There was no correlation of G 6 PD activities between erythrocytes and leukocytes both in 15 males and in all of 19 subjects.

The results mentioned above suggested that the enzyme activity in erythrocytes of G 6 PD variant(s) was obviously deficient in Guangdong. Leukocyte G 6 PD was also deficient significantly, but individual enzyme activities distributed widely and some were above the inferior limit of normal control. There was no parallel relationship of G 6 PD activities between two kinds of cells. G 6 PD activities in erythrocytes did not reflect G 6 PD activities in leukocytes.