

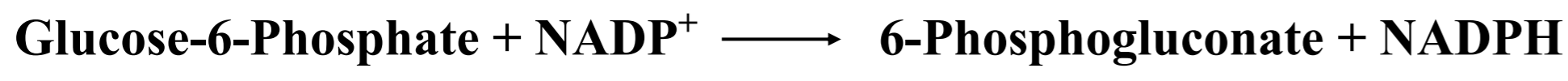
# Outcome of Newborn Glucose-6-Phosphate Dehydrogenase (G6PD) Screening Program in Taiwan

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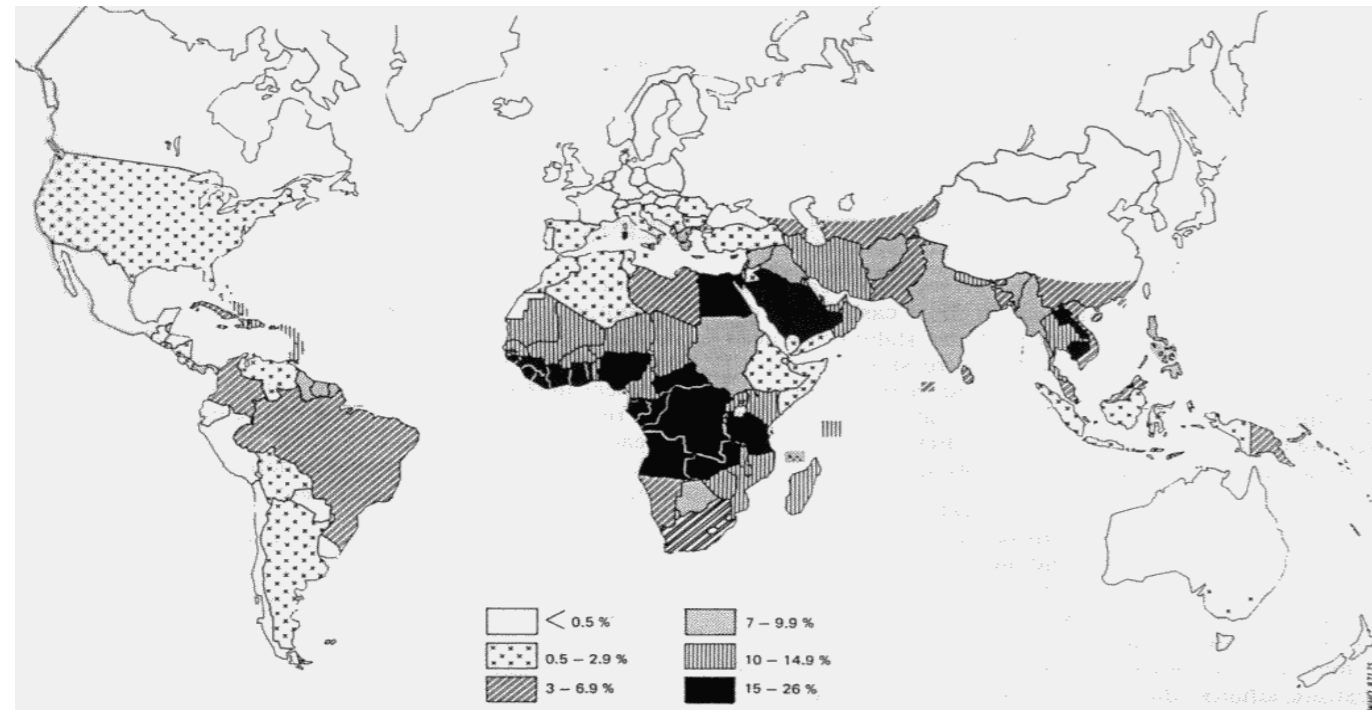
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## Introduction

### Glucose-6-Phosphate Dehydrogenase (G6PD) Deficiency



- X-linked recessive disorder (MIM 305900)
- Most common hemolytic disease in human
- 3.4% world population, 4.5M born annually with the deficiency



G6PD Deficiency Worldwide Distribution  
WHO working group. Bull WHO 1989;67:601

- Severe neonatal jaundice (NJ) triggered by environmental factors and/or medications is the major health impact of G6PD deficiency in newborns.
- Severe NJ may lead to kernicterus and cause death or permanent neurological damages.

## Severe Neonatal Jaundice in Taiwan before G6PD Screening

- **1973 - 1975: ( National Taiwan University Hospital )**
  - 30% of neonatal jaundice hospital admission were G6PD deficient
  - 15.8% mortality in those G6PD deficient neonates
  - 31.6% of them developed kernicterus

Chang MH et al. Acta Paed Sin 1977;18:1-10



- **Factors induced severe neonatal jaundice**
  - Naphthalene moth ball
  - Herbal medicines

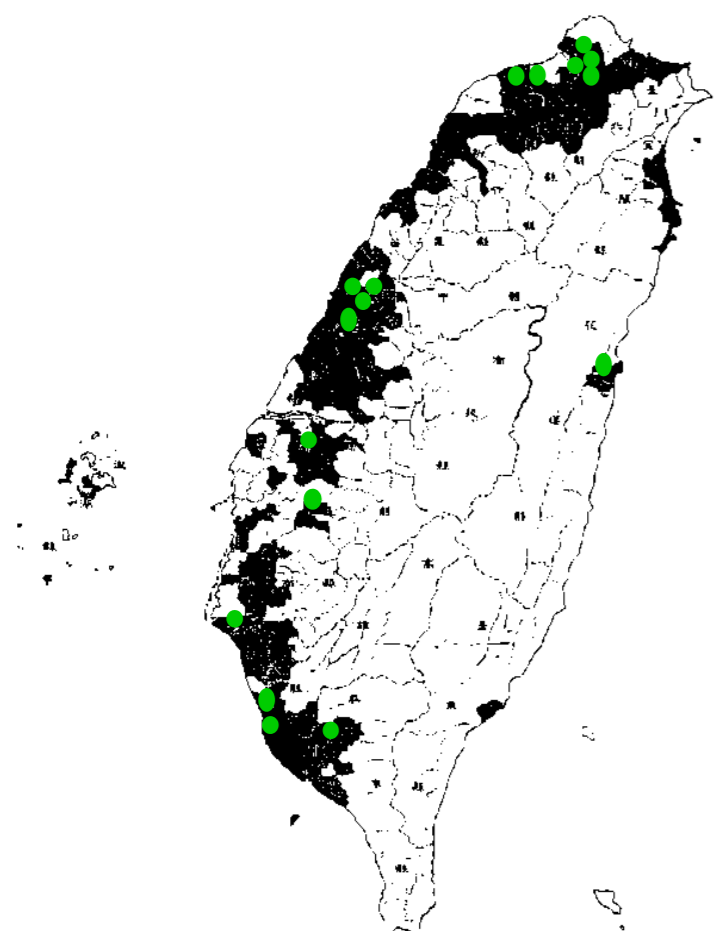
## Newborn G6PD Screening in Taiwan

- **Nationwide** routine G6PD screening program started in **1987**
  - Prevent severe NJ triggered by environmental factors and/or medications.
- **Preventive education** provided by the birthing facilities before discharge
  - Avoid contacting naphthalene
  - Don't give any medication to the newborn without prescription
  - Watch out for jaundice
- **3 Newborn Screening Centers**
  - Dried blood spots collected on day 2 ~ 5
  - Fluorescence spot test → Semi-quantitative test
  - > 97% of G6PD positive cases were informed before 7 days old
- **18 Referral Hospitals**
  - Quantitative G6PD confirmatory test
  - Clinical follow-up and genetic counseling
- **External Quality Assurance (EQA) Programs**
  - G6PD Quantitative Confirmatory Test since 1988
  - G6PD Blood Spot Screening Test since 1999



Chiang SH et al. Southern Asian J Trop Med Public Health 1999;30:72

### Geographical Distribution of the Participants in EQA Program for G6PD Confirmatory Test



- Participating laboratories (n = 18)
- Population density

## Results of Newborn G6PD Screening in Taiwan

- ~7 M newborns screened (1987 - 2017)
- Since **1996**, the effective coverage rate has reached **>99%**
- Screening results and efficiency (2013 ~ 2015)
  - Positive : ~ **2.5%**
  - Positive result notification before 7 days old : **97.4% ~ 99.0%**
  - Successful referred : **86 ~ 91%**
  - Confirmed : **80 ~ 92%**
- The overall incidence of G6PD deficiency in Taiwan : ~ **2%**
- **2000 - 2004: ( Veterans General Hospital medical center )** Yen HJ et al. Clin Neonatol 2005 ;12:1
  - Only **10** exchange transfusion (ET) required from **1,630** NJ admission, and **no mortality** was found ( ET/NJ = **6.13%** ).
  - Only **1** of exchange transfused had staphylococcal sepsis with G6PD deficiency

## Outcome of Newborn G6PD Screening in Taiwan, 2000 ~ 2010

- To access the outcome of newborn G6PD screening program, the patient data of those hospital admissions with NJ, after discharged from birthing facilities, between 2000 and 2010 was studied.
- The patient data were retrieved from the National Health Insurance Research Database (NHIRD), which covered >98% population of Taiwan, for analysis.
- Admitted NJ cases were identified as newborns who were hospitalized with NJ diagnostic code (ICD-9-CM, 774) and/or the procedure codes of phototherapy (ICD-9-CM, 99.83).

### Newborn Admitted to Hospital with NJ after Discharged from Birthing Facilities

Year	2000	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	Total
Birth No. <sup>1</sup>	305,312	260,354	247,530	227,070	216,419	205,854	204,459	204,414	198,733	191,310	166,886	2,428,341
Admitted NJ	812 (0.27%)	1,006 (0.39%)	1,080 (0.44%)	933 (0.41%)	1,049 (0.49%)	1,095 (0.53%)	1,170 (0.57%)	1,415 (0.69%)	1,451 (0.73%)	1,366 (0.72%)	1,405 (0.85%)	12,782 (0.53%)
ET <sup>2</sup>	3	5	6	6	2	2	-	-	-	2	1	27 (0.21%)

1. Data from "Department of Household Registration database, Ministry of Health and Welfare, Taiwan"
2. ET (Exchange Transfusion) cases were identified by the procedure codes of ICD-9-CM, 99.01

Although the cases of hospital admission with NJ after discharged from birthing facilities were increased slightly, the numbers of severe cases who needed to be treated with ET were dramatically **decreased to 0 ~ 2 cases per year** since 2004.

### Mortality and Severe Morbidity of Admitted NJ Newborns

Year	2000	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	Total
Birth No.	305,312	260,354	247,530	227,070	216,419	205,854	204,459	204,414	198,733	191,310	166,886	2,428,341
Admitted NJ	812	1,006	1,080	933	1,049	1,095	1,170	1,415	1,451	1,366	1,405	12,782
Mortality*	-	-	-	-	1 (0.09%)	-	-	2 (0.14%)	1 (0.07%)	3 (0.22%)	2 (0.14%)	9 (0.07%)
Kernicterus*	-	1 (0.10%)	-	-	1 (0.09%)	-	-	-	1 (0.07%)	1* (0.07%)	-	4 (0.03%)

\* Within 1 month of age

- None of the **dead cases** (most of them are premature babies) had **HHA** (Hereditary Hemolytic Anemia, which includes G6PD deficiency)
- Only **1** kernicterus case (\*) had **HHA**

## Conclusions & Discussion

- Newborn G6PD Screening Program in Taiwan (since **1987**)
  - Providing preventive education before discharge to all parents
  - > **97%** of G6PD positive cases were informed before 7 days old
  - The overall incidence of G6PD deficiency : ~ **2%**
- The admitted NJ newborns who need to be treated with **exchange transfusion** were dramatically decreased to **0 ~ 2 cases per year** since 2004 nationwide.
- The public health prevention program (including universal newborn G6PD screening) and effective clinical management in Taiwan almost **eliminated** all the "severe morbidity and mortality" caused by NJ in Taiwan.

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