



MyBaby & PKU

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1) MyBaby's diagnosis

- **newborn screening**
- **call from Sick Kids**
- **visit to PKU clinic at Sick Kids – test results**
- **admission to hospital – tests and consultations**
- **breastfeeding and formula for 6 months**



2) What is PKU ?

- not ----- allergy, contagious or life-threatening**
- metabolic genetic disorder**
- rare – 1 in 12, 000 or so; depends on location and ethnic group**
- first identified in early 1930s – Dr. Folling in Norway: persistent mom with two affected children**
- in next 10 years, discovered could not metabolize or break down phenylalanine (PHE), an amino acid in all proteins (building blocks – 10 aa in proteins)**
- deficient or lacking enzyme Phenylalanine Ammonia Hydroxylase**
- Phe to Tyrosine**
- Phe build up toxic to brain - esp. in first few years**
- severely to profoundly retarded, seizures, psychiatric and behaviour problems**
- thus, VERY SEVERE if untreated**
- early 1950s – Dr. Bickel in UK developed first diet to prevent this; persistent mom with 1 affected child**



- 1961 – Guthrie devised a PKU screening test - mass screening started and he traveled the world as an advocate. His son was disabled (unknown reason) and his niece was diagnosed with PKU**

- Universal newborn screening for PKU began in Ontario on June 1, 1965**



3) MyBaby's prognosis

- diagnosed and treatment started early**
- thus, normal growth, development, and learning**
- constant monitoring of blood levels of PHE to ensure within goal range**
- if fed foods medium or high in protein -- PHE levels rise --- brain damage**
- diet for life and keep track of what eating**
- good successes with other PKU children – future**
- no cure – only treatment through diet.....**



4) **MyBaby's care**

- **restrictive (low protein) diet – lots of foods have protein and thus, PHE**
 - **NO meat, fish, poultry, milk, eggs, cheese, legumes, nuts, soy**
 - **baked products – flour**
 - **1g or less – grocery store**
 - **currently eats ----- equivalents (PHE)**
 - **calories, vitamins, minerals need focus**
 - **weighed/measured food**
 - **will mostly eat fruits, veg, and lo protein products (pass around)**

- **medical formula (“Phenex”) – for her protein needs**
 - **“recipe” changes**
 - **(pass around)**

- **daily food logs – explain and pass around**

- **weekly blood tests (via mail) and review of results – (pass around)**
 - **goal range – between 200 and 400 PHE**

- **phone consultations with O's dietician every 1-2 weeks – adjusts diet and recipe/amount of formula**

- **illness**
 - **PKU clinic visits (now every 3 months) with dietician, nurse, PKU doctor – explain what happens**
 - **lots of love! No PITY**



5) Working together – how **YourDaycare can help us care for **MyBaby****

- **records of what eaten – not as much detail**
- **formula – record amount – will continue in preschool room etc.**
- **teach her YES and NO foods**
- **communication with us – any questions etc.**
- **if not sure.....ask!**
- **information posted for staff who aren't regular**
- **we will supply food – explain**
- **much of BLUE WAVE menu – won't work**
- **(pass around books)**

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