Robin and I Explain PKU

to children who need a very important diet

and

to their friends and relatives of all ages

Foreword

Jane writes,
Our son Bill was diagnosed with phenylketonuria (PKU) at the age of eleven days in December 1968. He has remained on his PKU diet ever since and from a young age we have assured him that this diet should never stop him doing anything. He is now an economist, with an Honours degree from Adelaide University and a Masters from Queen's University, Kingston, Ontario, Canada. He and his wife Jane, who is also an economist, have two daughters. A highlight of Bill's career was working with Korean PKU children and their parents during his three year posting in Seoul, from 2002 to 2005.

I met six year old Caroline Thorpe, who has PKU, in 1978 when I became the teacher librarian at her school. In response to staff requests for information about PKU I wrote this book. It reflects the experiences of our family, including our daughter Susie, who does not have PKU. My medical husband Bob helped simplify scientific background material so that it met the needs of adults, and particularly grandparents. Caroline's response was a delight.

Caroline explains,
As a child I often wrote and drew my own books, some made out of Dad's old computer paper which he would bring home for me to draw on. So I guess a lot of my interests blossomed there. It was quite natural for me to draw the pictures for this book. Originally I saw the copy with only the words in it and being a kid I wanted to have a bit of a jot on the paper because of the pictures going through my mind when I read it. Simple as that. I did it in pencil, then people saw and liked my drawings and I was asked to elaborate on them with colour. It was a bunch of fun! I hope many more people find this book as enjoyable as I have.

Our book was first published in black and white by the Child, Adolescent and Family Health Service, Adelaide as a contribution to Hidden Disabilities Week in the International Year of the Disabled, 1981.

We warmly thank Virginia Schuett, editor of National PKU News, for making it available on www.pkunews.org This has also enabled us to share it in colour for the first time.

Jane Brummitt and Caroline Thorpe,
Adelaide, 2009.

Caroline Thorpe aged eight, 1981.

Bill Brummitt sharing koala and kangaroo stickers with PKU children in Seoul, South Korea. *The Korea Herald*, October 12, 2004.

First published in Adelaide, South Australia
by Child, Adolescent and Family Health Service, 1981.

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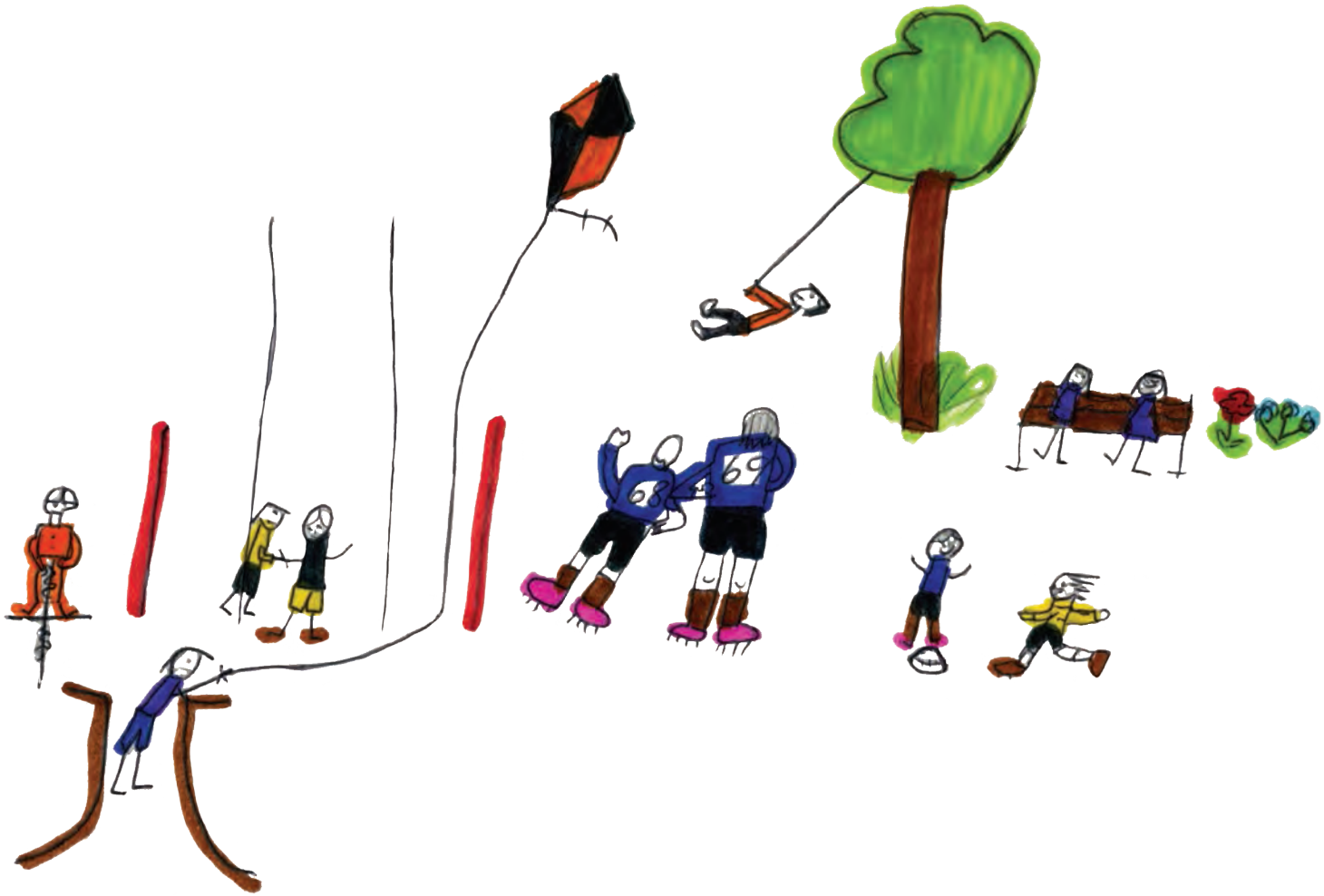
Robin and I Explain PKU

by
Jane Brummitt

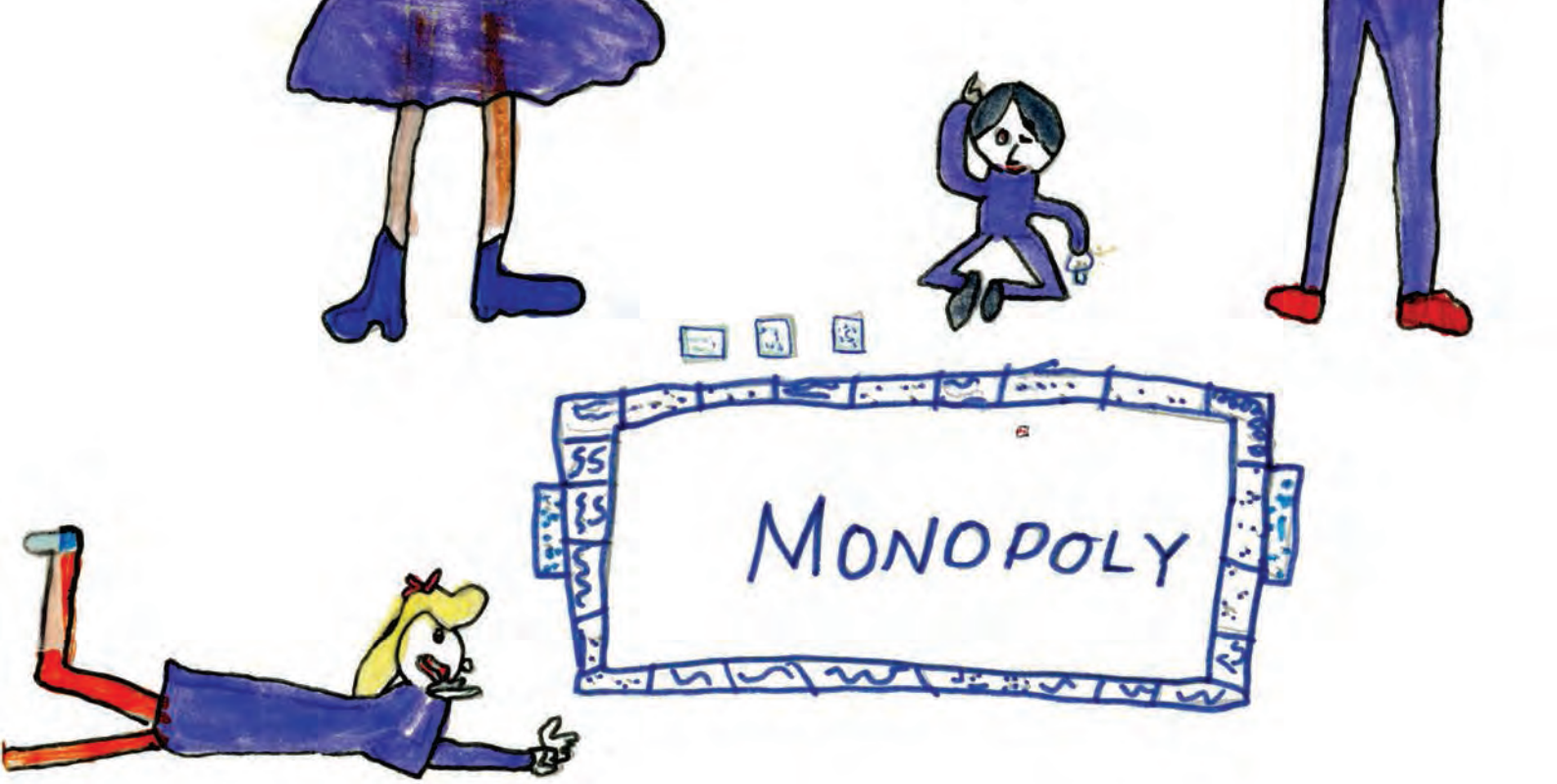
with illustrations by
Caroline Thorpe

medical consultant
Robert Brummitt

Robin and I,
And others too,

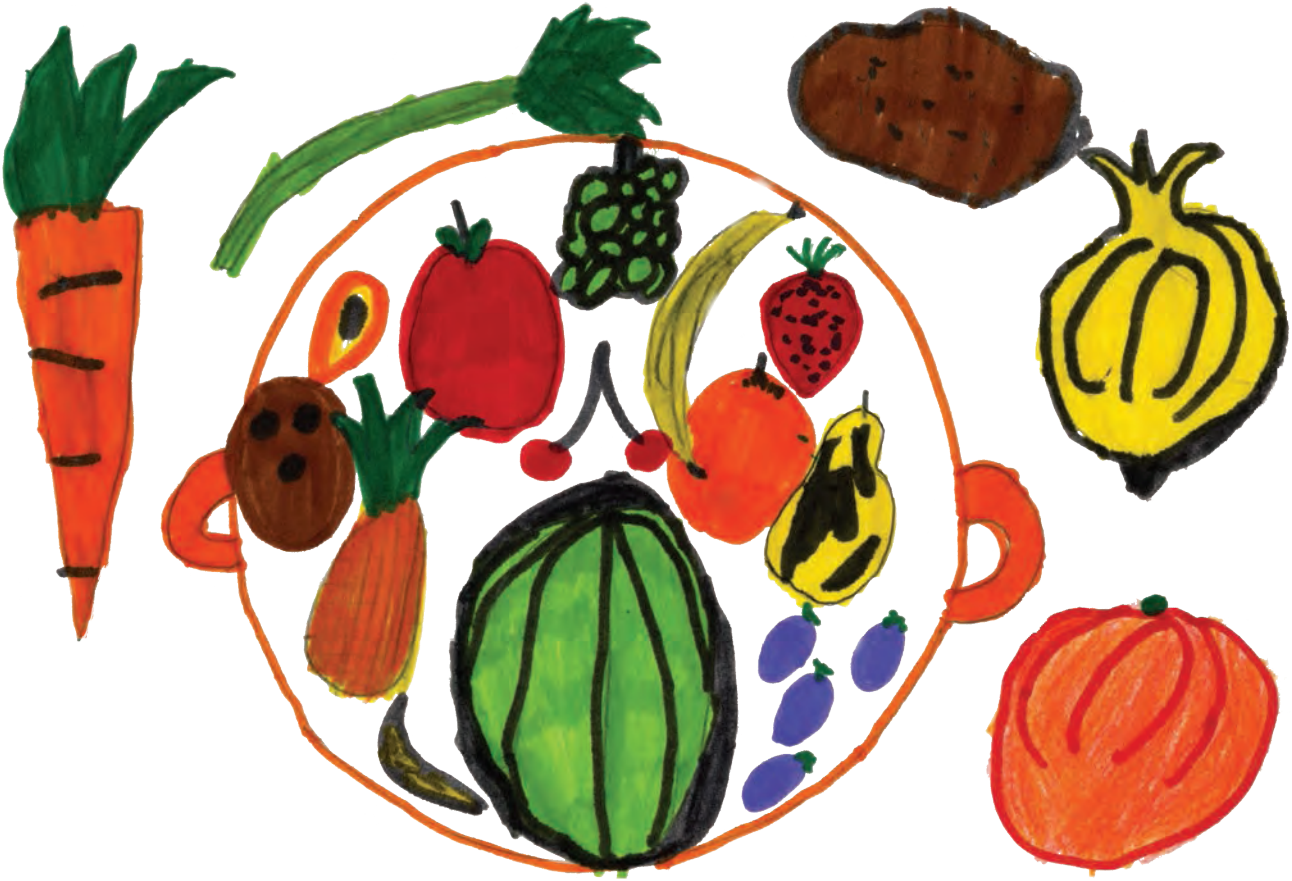


Are like anyone else,
But have PKU.



We love playing games
And we love going out,

And friends soon learn
What our diet is about.



The food right for us
Is the food we must eat –

rice

spaghetti

party patties

Pizza

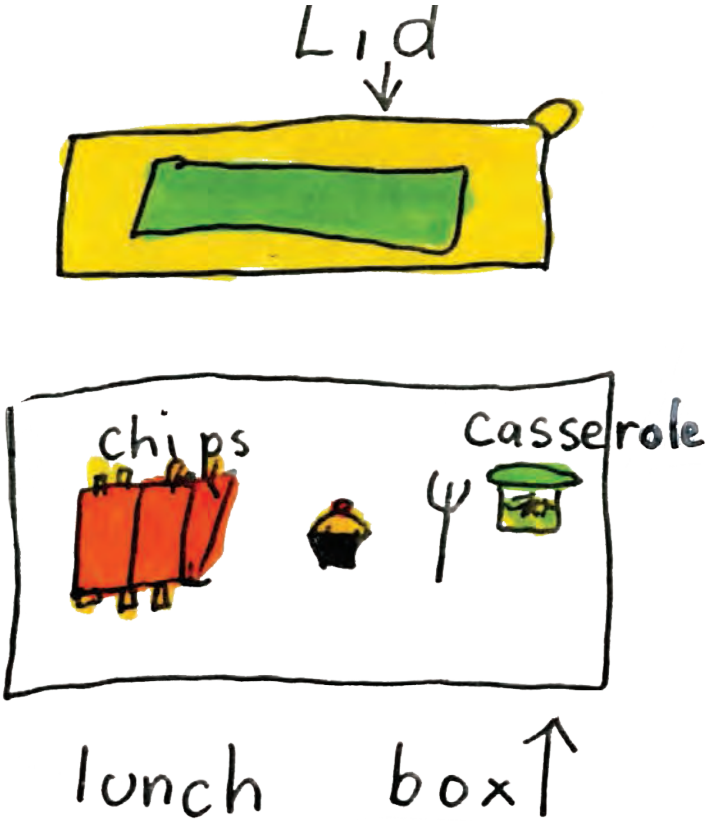
Agar agar
Jelly Mouse

Casserole

treat

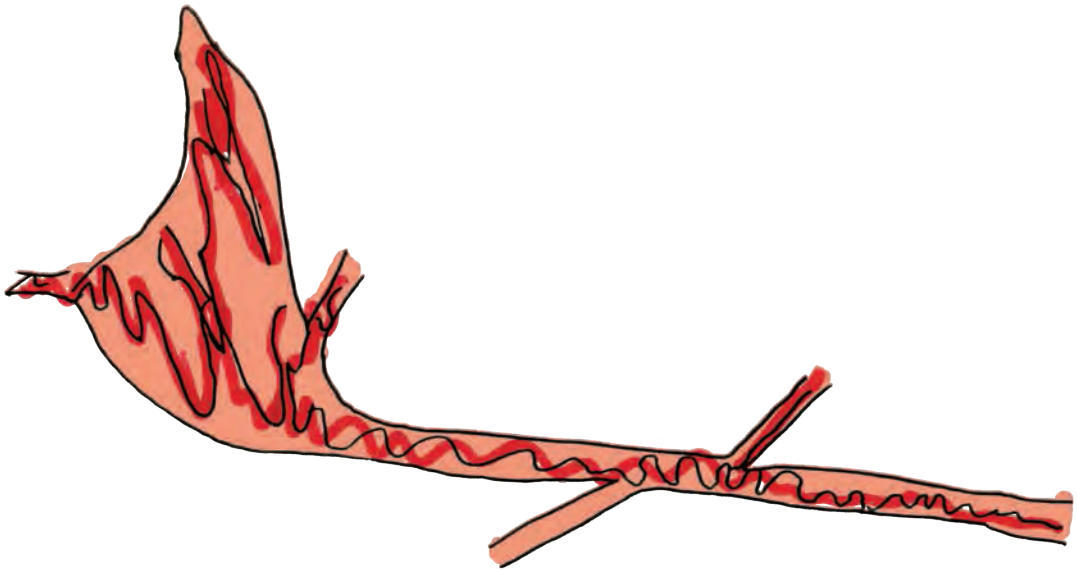
sweet!

Fruit, veggies and 'specials',
And sometimes a treat!



Some of our specials
Come travelling with us –

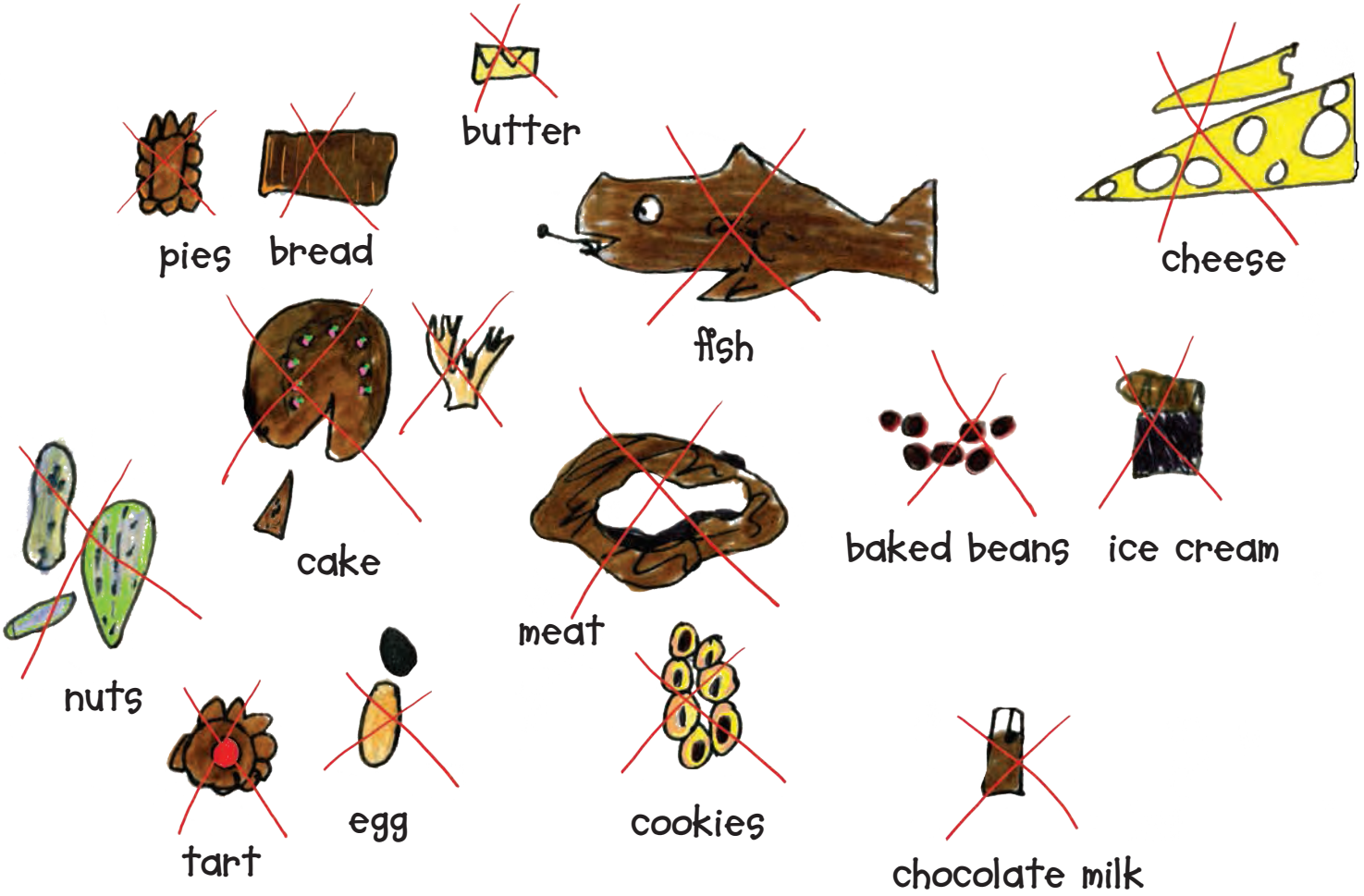
And it's never, (well rarely!)
We make any fuss.



Our blood knows some secrets
Most people can't see,

So it needs to be checked
Quite reg-u-lar-ly.

And daily we need
A magical potion,

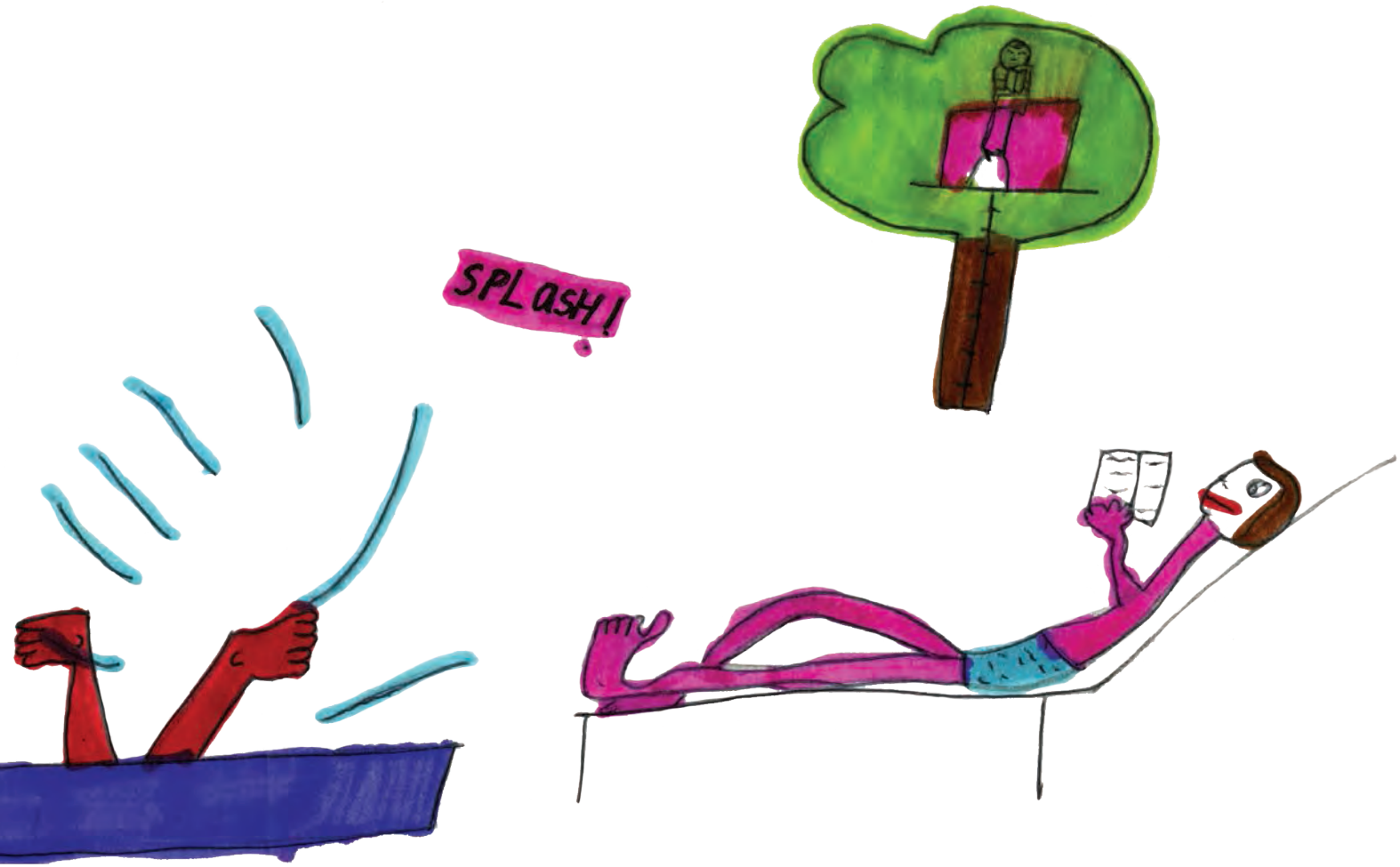


Instead of those foods
Which cause blood test commotion.

Our families and friends
Are proud of us all –

Whether we're short,
Or middling or tall.

So! Robin and I
Hope we've helped you, too,



To understand something
Of PKU.

TO ANSWER SOME OF THE QUESTIONS OFTEN ASKED ABOUT PKU,

further information is included in the following pages

What is PKU ?

How is it diagnosed ?

Why are some children born with PKU ?

What happens if PKU children are not started on this special diet ?

Why is the diet ‘special’?

Which ordinary foods do PKU children eat?

What is the magical potion ?

What are ‘specials’ ?

WHAT IS PKU ?

It is the abbreviation for a rare condition known as Phenylketonuria, which is treated by a special diet.

It is caused by the absence of just one particular enzyme from many thousands that are present in each of our bodies.

(Enzymes speed the change of one substance into another in the body's 'chemical factory'.)

The food we eat every day contains protein, fats and carbohydrates. Only the protein part causes trouble in PKU.

An important part of protein is called phenylalanine, which is quickly turned into useful chemicals by the body – except in people born with PKU, in whom the enzyme necessary to make this change is the one that is missing.

HOW IS PKU DIAGNOSED?

It is diagnosed by a blood test done around the fifth day of life.

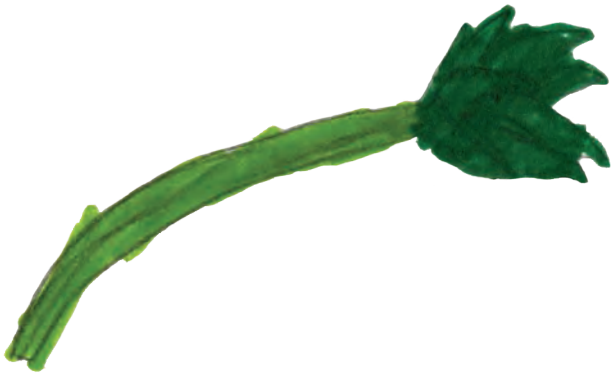
Approximately one in ten thousand babies will have a ‘positive’ result and will need to be placed on a special diet.

WHY ARE SOME CHILDREN BORN WITH PKU?

For a child to be born with PKU, both parents will have carried a 'hidden' gene for it.

About one person in fifty has the hidden gene for PKU. This means that on average, about one in every 2,500 couples will both have the hidden gene.

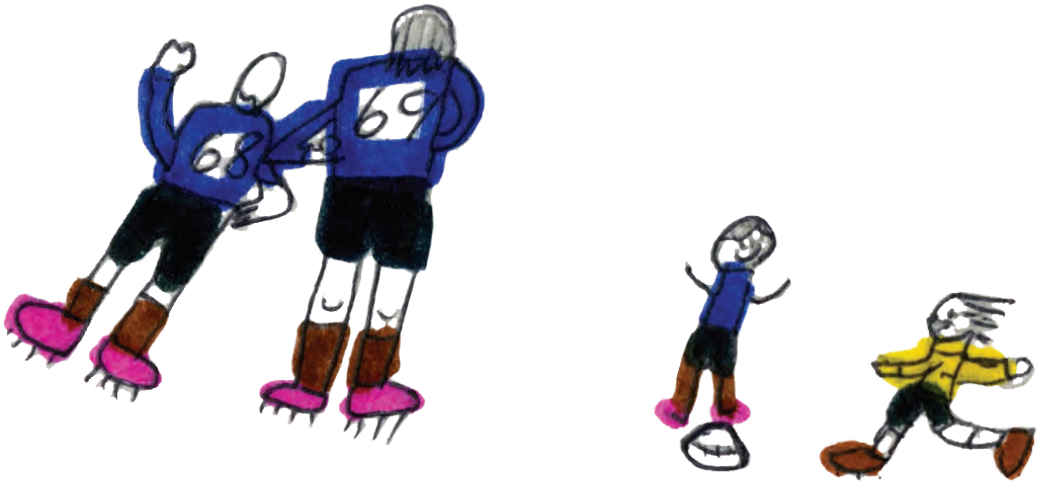
Though parents of PKU children do not have the condition themselves, each time they have a baby, there is a one in four chance that it will have PKU.



WHAT HAPPENS IF PKU CHILDREN ARE NOT STARTED ON THIS SPECIAL DIET?

If babies with PKU are not started on a special diet in the first two weeks of life, their blood level of phenylalanine gets too high and this starts to cause irreversible damage to their brains and bodies.

For well over forty years now, children who were diagnosed in the first fortnight of life and put on to the PKU diet have thrived and have not experienced this damage.



WHY IS THE DIET SPECIAL?

It is 'special' because it is low in protein. At first the baby is given a milk substitute instead of the milk it has had since it was born. As well as this, the baby will need a carefully regulated portion of breast milk or ordinary milk. This will be worked out by a dietitian, and will change as the baby grows.

Regular blood tests, exactly the same as the very first one, will continue to be done at home and sent to the hospital laboratory. The levels of phenylalanine will be checked to make sure they are neither too high nor too low. This means that the food eaten must be appropriate, so that there is no 'blood test commotion'.

PKU babies progress to solid foods at the same stages as other children, but they must not have high protein food. So they cannot eat meats, poultry, fish, eggs, milk, cheese, gelatine, chocolate, nuts or high protein vegetarian food, unless specially allowed by the dietitian.

WHICH ORDINARY FOODS DO PKU CHILDREN EAT?

They eat most fruit and vegetables and learn from an amazingly young age just which foods are right for them.

Parents will have lists of the other foods each individual is allowed. They will also have excellent recipe books. As children grow older they will gradually realise that they are caring for themselves properly by eating the right food.

WHAT IS THE 'MAGICAL POTION'?

It is a food mixture without the phenylalanine which cannot be tolerated. PKU children need this 'magical potion' for growing, just as other children need protein foods.

Others in the family, and friends too, need to understand its importance, just like the big sister who created the term 'magical potion' to describe the protein substitute her little brother needed every day.

People with PKU need to have both their ordinary foods and their protein substitute, or 'magical potion', every day.

WHAT ARE 'SPECIALS'?

The 'specials' PKU children need include low protein bread, flour and spaghetti etc. They can take these on holidays with the family, grandparents, friends etc. and to barbecues, hikes and camps.

PKU children often bring 'treats' of their own to parties - special pizzas or cakes for example - and they will look forward to other food they are allowed, party drinks and party fun! So they need never be left out of outings or parties because of their diet.

Afterword

This book was written as a tribute to the pioneers of early diagnosis and treatment of PKU at the Adelaide Children's' Hospital in the mid 1960s, namely

- **Dr John Covernton,** Medical Practitioner, who recognised the urgency of implementing the Guthrie blood test on newborn babies in order to prevent irreversible mental retardation, and made South Australia the first state in Australia to test all newborn babies.

- **Geoffrey Hill,** Biochemist, who (with the aid of volunteers) established the testing programme in the laboratory and the continued monitoring of children who had been diagnosed.

- **Alison Morrissey,** Registered Nurse in charge of the Diet Kitchen, who researched and managed the dietary treatment for children with PKU.

We also acknowledge the outstanding American, Dr Robert Guthrie, who not only developed the Guthrie screening test for PKU in 1961 but also promoted it worldwide. This has enabled the prevention of severe mental retardation in thousands of children and has allowed them to lead normal lives.

A badge given by Dr Guthrie to Bob and Jane Brummitt.