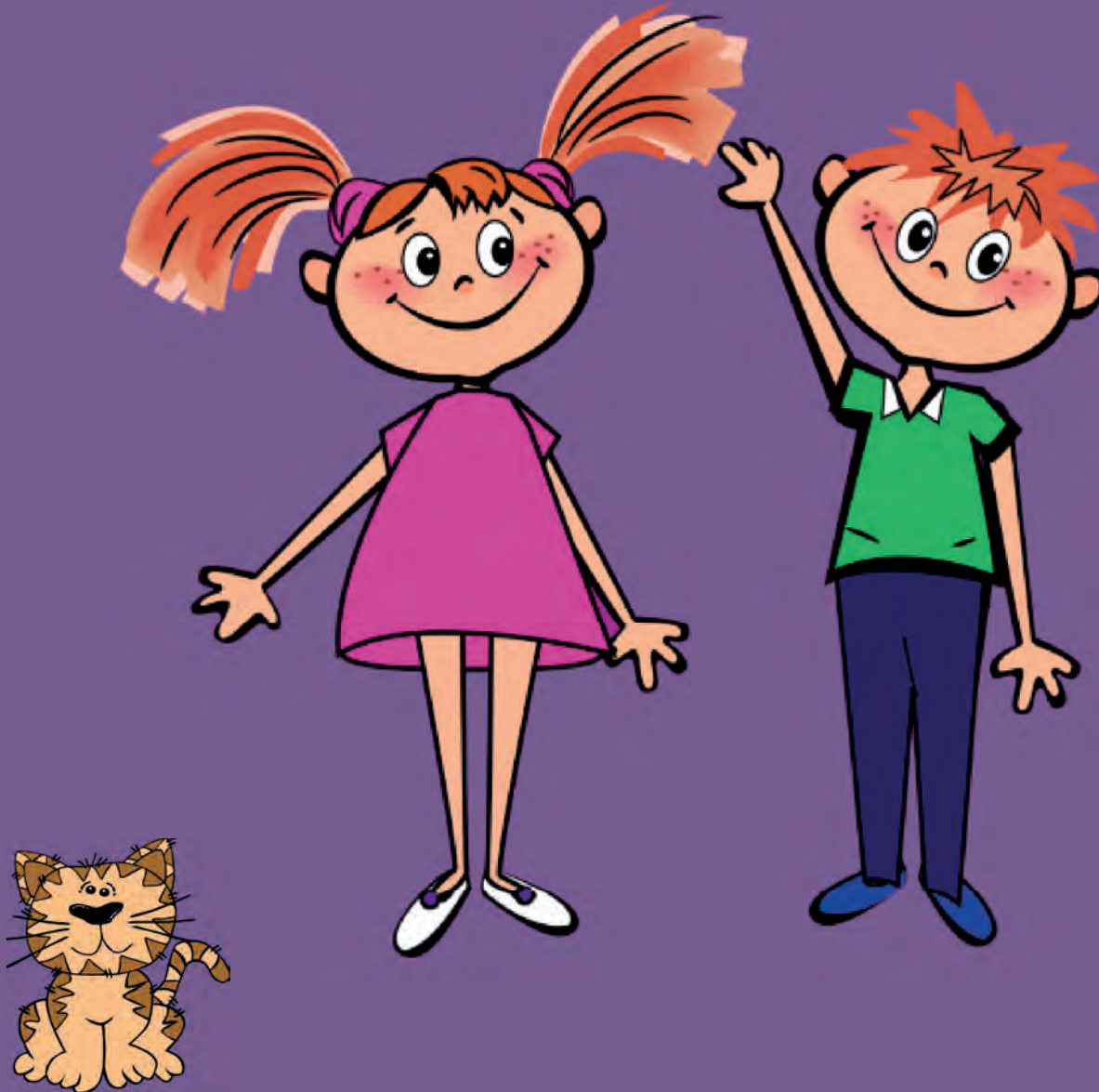


# Are You an Alpha Too?



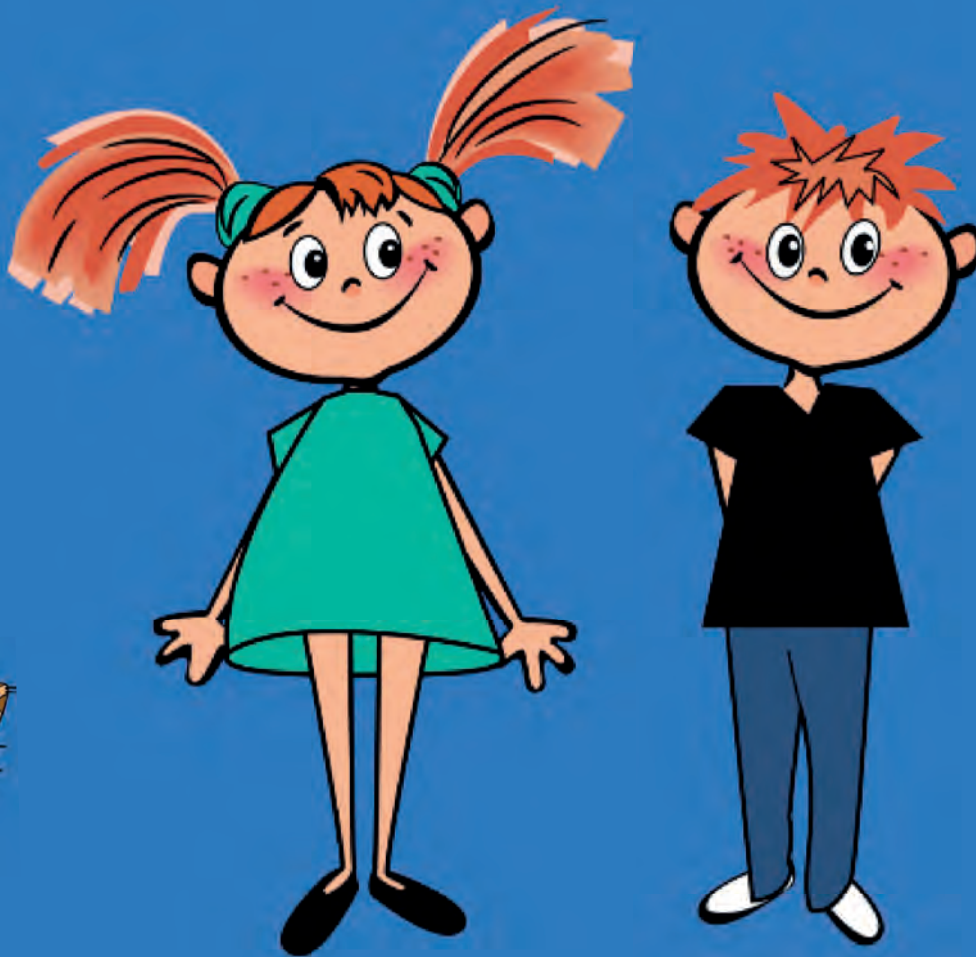
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A children's guide to alpha 1 antitrypsin deficiency

# Meet Lucy and Tom

We would like you to meet Lucy and Tom [and Tigger]. They also have a baby brother called Ben.

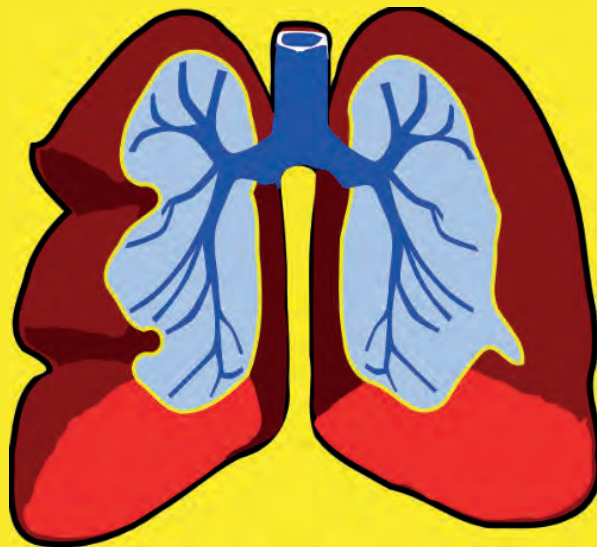
They are going to help us to tell you all about what it is like to be an alpha.



Alpha 1 antitrypsin deficiency is quite hard to say. It means that we do not have enough of something called alpha 1 antitrypsin in our blood. AAT is a short way to say alpha 1 antitrypsin, so we will call it that because it is easier to remember.

We need AAT to help keep our bodies to stay fit and healthy. It is especially important for the organs we breathe with. These are called lungs. AAT helps to protect them when we breathe in germs and other bad things like smoke. Because we do not have enough AAT in our blood, the bad things that we breathe in can hurt our lungs.

Your lungs are in your chest. If you put your hands flat on your chest you can feel them blow up like balloons when you breathe in, then go down again when you breathe out. They look like this.



AAT is made in another organ in your body called the liver. Your liver is below your lungs near your tummy. It looks like this.



When the liver has made the AAT it goes into the blood. Then the heart pumps it all round the body through little tubes called blood vessels. When it reaches the lungs it helps to keep them healthy by coating them with a protective layer. This stops the bad things from damaging the lungs.

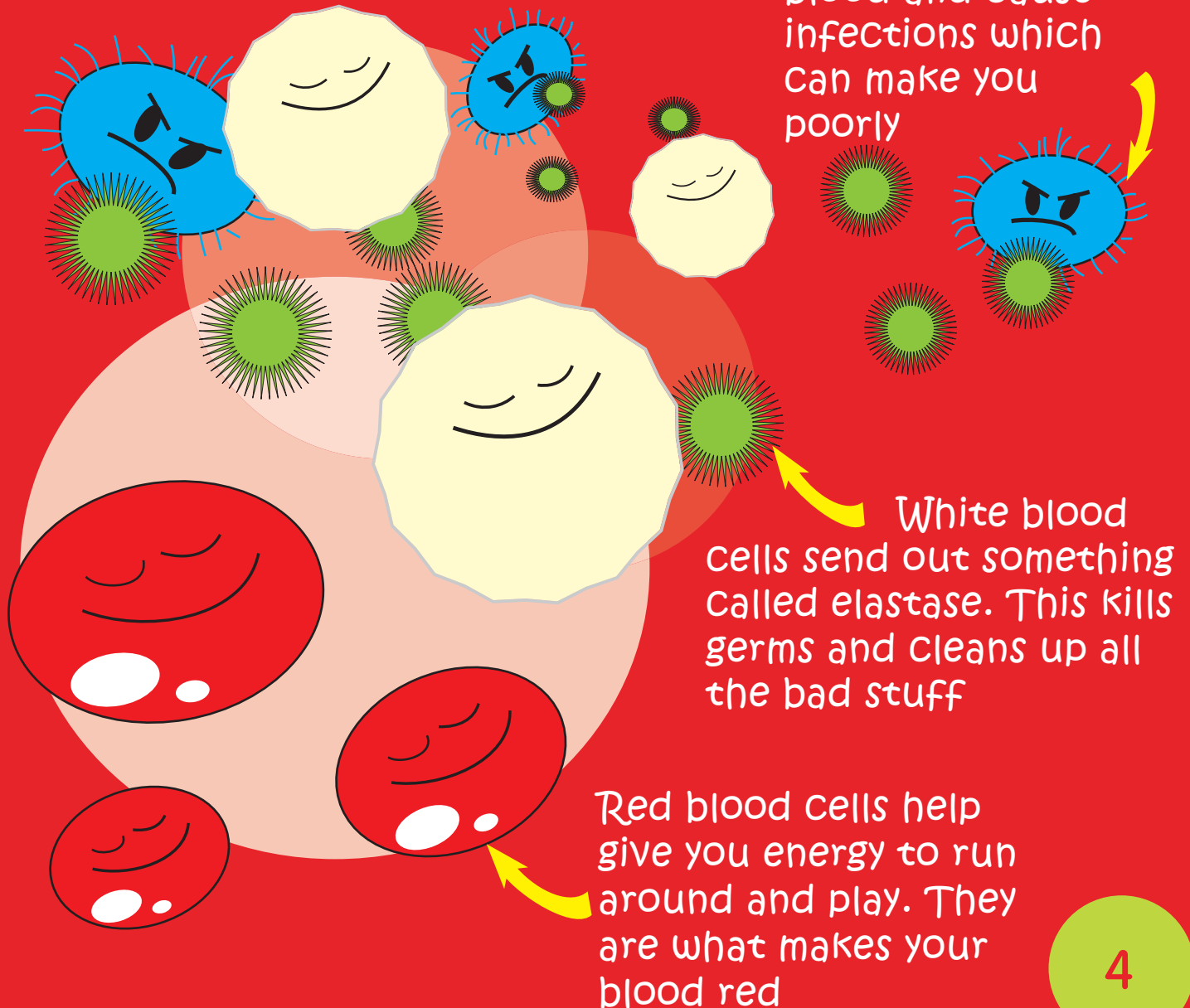
Because we are alphas the AAT that we make is not quite right. It is a funny shape and it gets stuck in our livers. Most of it cannot get out into our blood which means there is not enough to look after our lungs properly.



# Tiny things in your blood

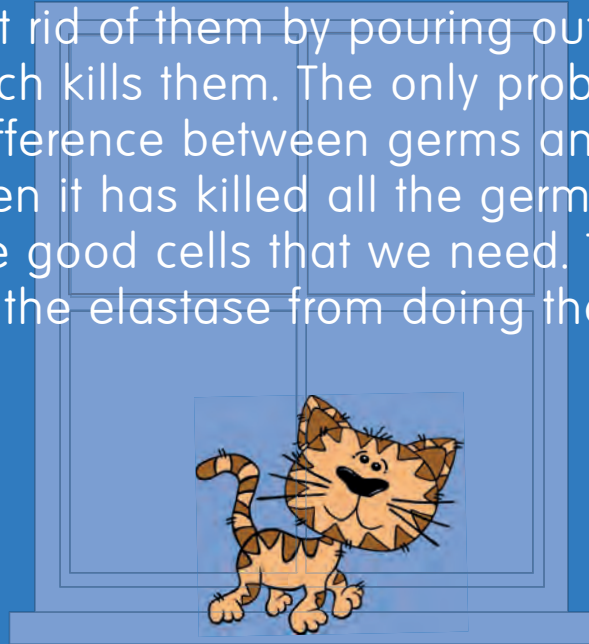
These are so small you can only see them with a microscope

Germs get into your blood and cause infections which can make you poorly





When we are poorly with coughs and colds, it is because we have caught germs from someone else. When germs get into our bodies they make can us feel ill. Our white blood cells help us, they find the germs and get rid of them by pouring out something called elastase which kills them. The only problem is that elastase cannot tell the difference between germs and the good cells in our lungs. So when it has killed all the germs and bad stuff, it will try to attack the good cells that we need. The AAT coating in the lungs stops the elastase from doing that.



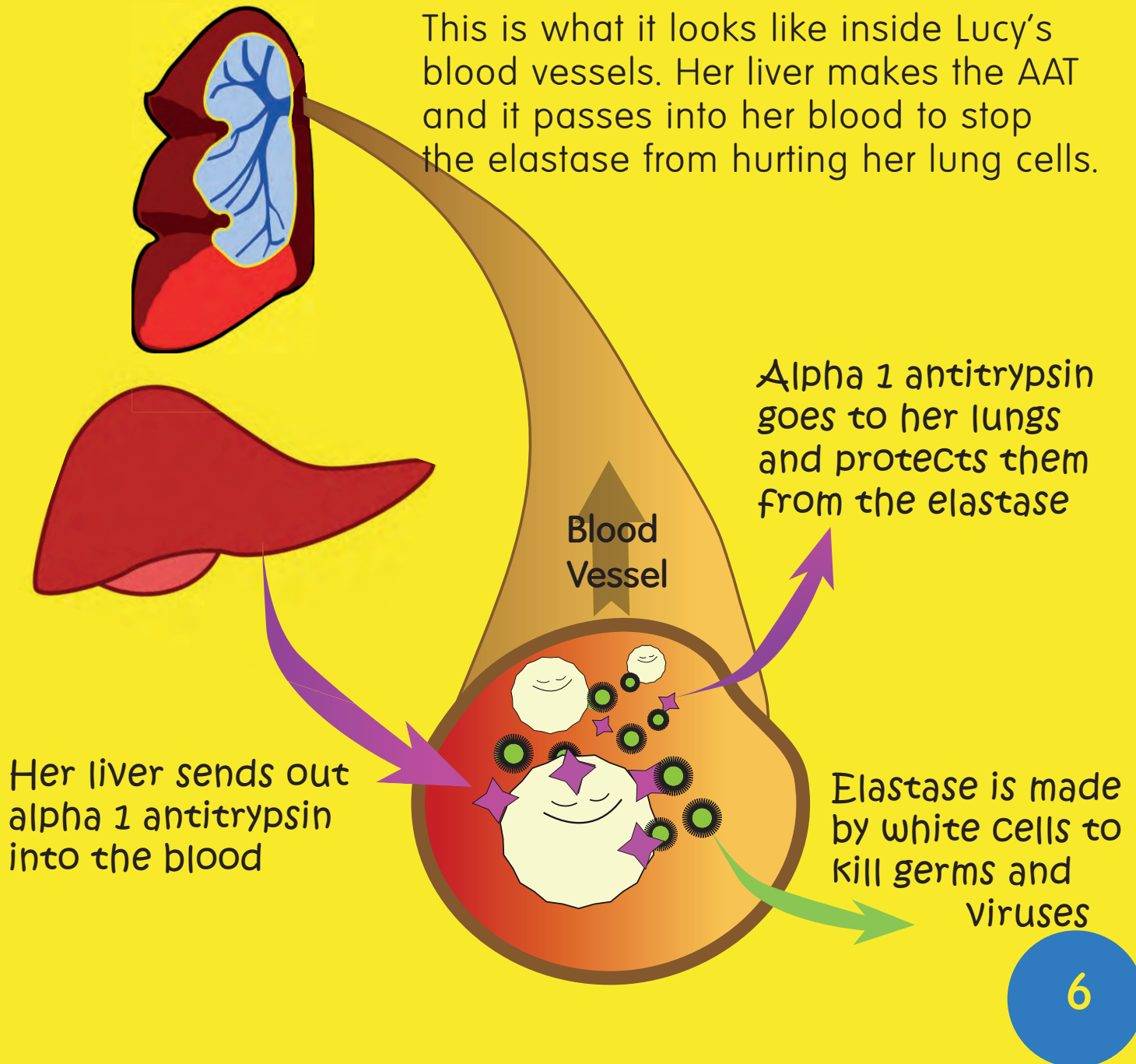
Lucy has less AAT than someone who doesn't have AAT deficiency but she does have enough to keep her lungs healthy.

She is called an alpha carrier. Lucy and Tom's mum and dad are carriers too. Lucy should be fine if she looks after herself and never smokes cigarettes when she grows up.



# Inside Lucy's blood vessels

This is what it looks like inside Lucy's blood vessels. Her liver makes the AAT and it passes into her blood to stop the elastase from hurting her lung cells.



Tom's blood has a lot less AAT in it than Lucy's. He is called an alpha. Because his AAT is a funny shape it gets stuck in his liver and so not much of it gets into his blood to help his lungs.

He has to be careful when he gets a bad cough because the elastase in his blood can hurt his lungs. He will need to take special medicines called antibiotics when he has a cough. They will kill off a lot of the germs so that the white cells don't have to make so much elastase.



It is very important that Tom has an injection every year before the bad Winter weather comes along. This will help to stop him getting the 'flu. 'Flu is like a really bad cold. Injections are not very nice but they only hurt for a little while and that is better than being poorly for a long time.

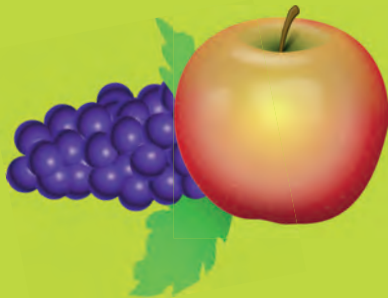
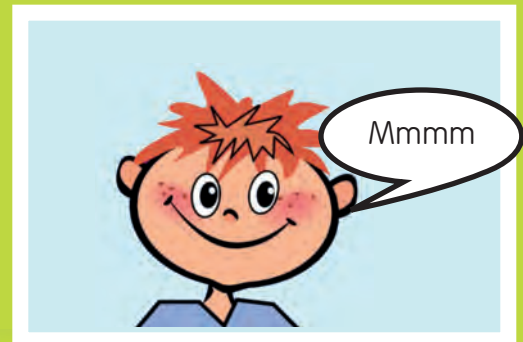
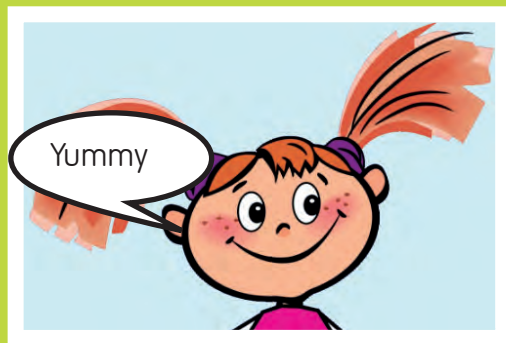
Tom will always have to be careful about breathing in things that might hurt his lungs.



Like Lucy and you, he must never smoke cigarettes. They will damage his lungs badly and could make him very poorly when he gets older. Sometimes older people with AAT deficiency who have smoked get lung disease and they cannot breathe properly. Do you have someone in your family like that?

It would be best if Tom stays away from any sort of smoke. If there are any people in your family who smoke cigarettes, they must not smoke them anywhere near you.

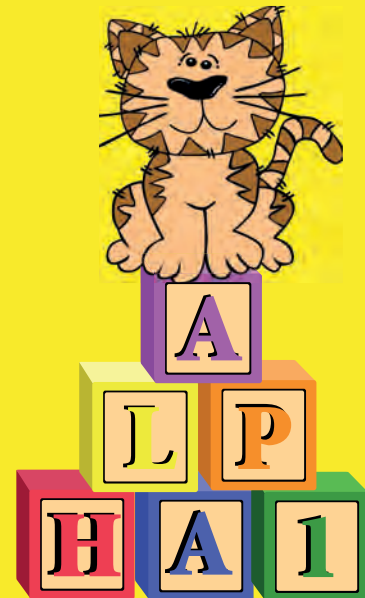
Tom and Lucy eat lots of things that are good for them. They never used to like fruit and vegetables but they do now and they eat lots. They can still have other things that they really like as well but they do not have things like burgers and chips too often.



# Meet Lucy and Tom's baby brother Ben

Ben is an alpha like Tom, he doesn't have much AAT in his blood. Poor little Ben has been quite poorly. Most children who do not have enough AAT in their blood do not have any problems when they are growing up. Sometimes though, an alpha baby can be born with a liver that isn't working properly.

That is what happened to Ben. When he was born Ben's skin looked yellow. The doctors said it was called jaundice and that he looked like that because all the funny shaped AAT has got stuck in his liver and it wasn't working properly.



Sometimes Ben's tummy feels sore and it makes him feel poorly. Ben has to have special foods to eat and special milk to drink in his bottle. These will help him to grow big and strong. If he did not have the special foods he would not be able to grow properly.

Ben sees a lot of people who help his family to look after him. Doctors and nurses make sure he is growing well and eating properly. He often has to go to the hospital too. Special doctors there examine Ben and then chat to Mum or Dad and tell them how well he is doing. They will also talk about anything else that can be done to help Ben.



Sometimes they have to do special tests or look inside Ben with a special machine called an ultrasound scanner. It scans over his tummy and shows pictures of what Ben's liver looks like under his skin. The scan does not hurt but sometimes Ben wriggles about so much that it makes it hard to see the pictures.



A lot of babies like Ben will get better as they get older and their livers will start to work properly. We hope Ben's will too.

## How do you feel?

Lucy and Tom help to look after Ben and they play with him when he feels well. They do get a bit fed up sometimes though because their mum and dad spend a lot more time with Ben when he is poorly. Ben cannot help being poorly but sometimes it seems like they have no time for Lucy and Tom.

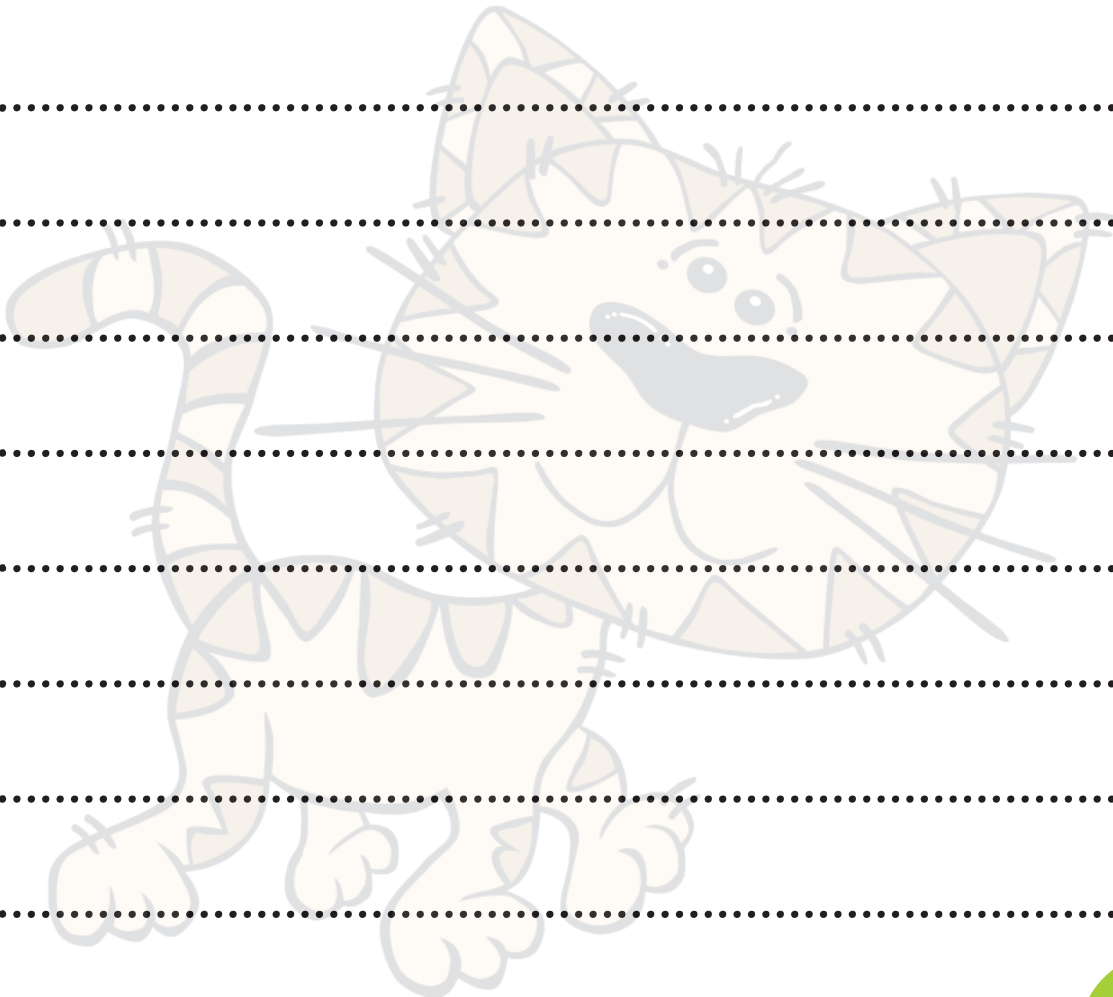
Perhaps you have felt the same if you have a brother or sister who is more poorly than you are. It is normal to feel like that. Mums and dads love all their children but when someone is poorly they need their mum and dad more for a little while.

Do you feel upset or worried about yourself, or a brother or sister? Maybe it's Mum or Dad who are poorly and you worry about them. If you do you should let someone know how you are feeling. If you do not want to upset Mum and Dad, maybe you could talk to Nana or Granddad. It is much better to tell someone how you feel than to keep it hidden inside. They will help you to understand what is happening.



## Your questions

This page is for you to write down anything that you may be worried about, or any questions you want to ask your family or doctor.





# About you

This booklet is yours to help you to understand a little bit about being an alpha and to explain some of the things that may happen to you or to other family members.

You can fill in your name and other things here.

my first name:

.....

my age:

.....

my school:

.....

my teacher is called:

.....

my hospital:

.....

my doctor is called:

.....

my name is  
Tigger



# A picture for you to colour



Lucy has a friend whose mum is poorly because of AAT deficiency.  
This is a poem about her.

My mum looks like a spaceman,  
With tubes to help her breathe.  
She helps me with my homework,  
Except when I cough and sneeze.  
She can't play running games,  
Or move as quick you see.  
When she tries, she starts to cry  
And needs a hug from me.  
Mum tries to make the best of things  
And does just what she can.  
She knows we always love her,  
She's our very special mum.



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Booklet written and produced by  
Alpha-1 UK Support Group ©  
With special thanks to Joe for the poem  
[www.alpha1.org.uk](http://www.alpha1.org.uk)

Special thanks also to  
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Mum's got this condition  
That stops her doing things.  
I help her with the shopping  
And do the ironing.  
Mum hasn't got enough AAT  
To keep her fit and well.  
It's only when she gasps for breath  
That you can really tell.  
I wish there was a medicine  
That really would make her well.

AAT means living carefully  
Avoiding smoke, fumes and things.  
Staying away from infections  
With the problems that they bring.

(Joe Lyons 2011)