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Cancer Council Victoria

FAP

Familial Adenomatous Polyposis

Information for families

Familial adenomatous polyposis (FAP)

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Introduction

This booklet has been written for people who have, or are at risk of, familial adenomatous polyposis (often simply called familial polyposis or FAP).

It tells you what FAP is and what can be done about it.

Two points are emphasised. First, many people with FAP live long and active lives after treatment, despite some inconveniences. There is much that can be done for FAP. Second, if you know you have FAP, or if you are at risk, it is vital that you have regular check-ups with your doctor. The check-ups should start when you are in your mid teens. This gives the best possible chance of successful treatment.

You may like to pass this booklet on to your family and friends for their information. This booklet does not need to be read all at once, but can be read in sections according to your needs or interest. The words in **bold** are explained in the glossary.

*** Are you reading this for someone who does not understand English? Tell them about the Multilingual Cancer Information Line. See the inside back cover for details.**

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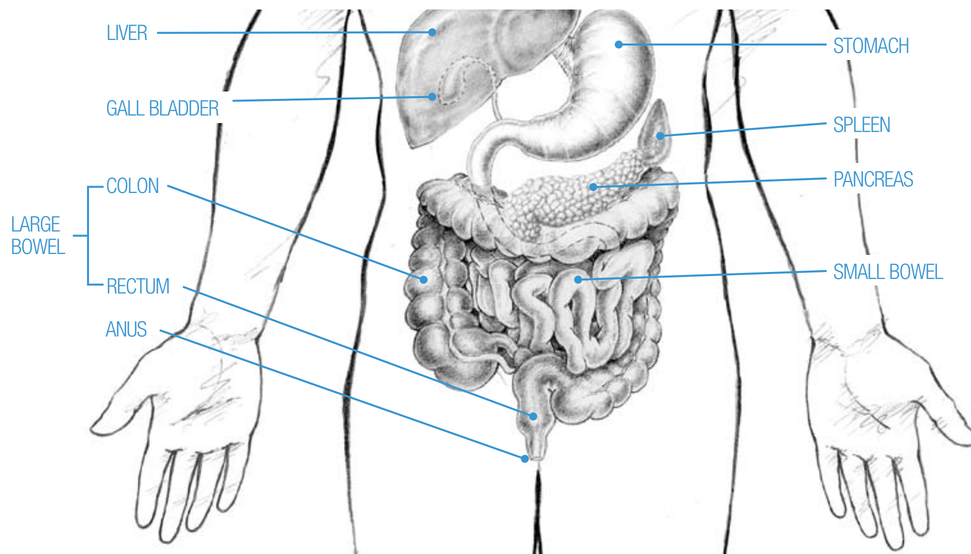
The bowel

The bowel is the longest part of the digestive system (the 'gut').

The digestive system is the long tube that runs from the back of the mouth, forms the stomach and **bowel**, then ends at the anus. It winds around inside the body. Food passes through it and is broken down and absorbed. The waste products are passed out as bowel motions.

The bowel is made up of two sections. The small bowel (the **duodenum**, **jejunum** and **ileum**) is where food is absorbed. This leads into the large bowel, where only water and salts are absorbed. The **large bowel** has two parts: the **colon**, which is about one and a half metres long, and the **rectum**, which is about fifteen centimetres long. The rectum leads to the outside of the body through the **anus**.

The digestive system



Familial adenomatous polyposis

Familial adenomatous polyposis is pronounced fam-il-ee-al ad-en-oh-mat-us poly-poh-sis. It is often shortened to familial polyposis or FAP.

- FAP is *familial*, that is, it runs in families. If one of your parents has FAP, you may be at risk. If one of your brothers or sisters has FAP, you may also be at risk.
- The polyps in FAP are *adenomatous*, which means they are the type that can develop into cancer. If the polyps are not treated, a **bowel** cancer will develop in one or more of them.
- *Polyposis* means a lot of polyps in the bowel (see below). Polyps are mushroom-like growths.

What causes FAP?

FAP is caused by a change in a **gene** called the **adenomatosis polyposis coli** (APC) gene, which in this booklet is called the 'FAP gene'.

Genes carry the instructions that control how our bodies grow, develop and work. Each cell of the body carries the whole 'genetic plan' for the body-made up of tens of thousands of different genes.

Among its important functions, the FAP gene helps control the way the lining of the **bowel** develops. Everybody has this gene, but only people with a change in the gene (a **mutation**) get FAP. Once this change has occurred, it may be passed from parent to child (that is, the child may inherit the change from their parent).

Everyone has two copies of each gene: one from their father, and the other from their mother. Usually the two genes act in the same way, but if they contain different instructions, one gene may dominate the other. This is what happens with FAP. If you inherit a changed FAP

gene (or if the gene change occurs in your body), the changed gene will dominate the other gene, and you will develop FAP.

Who is at risk of FAP?

If you have a parent with FAP, you have a 50:50 chance (or one chance in two) of having FAP. If you have a brother or sister with FAP, you may also be at risk of FAP. The risk is the same for men and women.

If you have FAP, each of your children has a 50:50 chance of inheriting the changed FAP **gene** from you. Remember that because genes are inherited in pairs, if you have FAP you will have one copy of the altered FAP gene and one working FAP gene. When you have a child you will pass on one of these copies, but not both. It's like a toss of a coin whether the changed copy or the working copy is passed on.

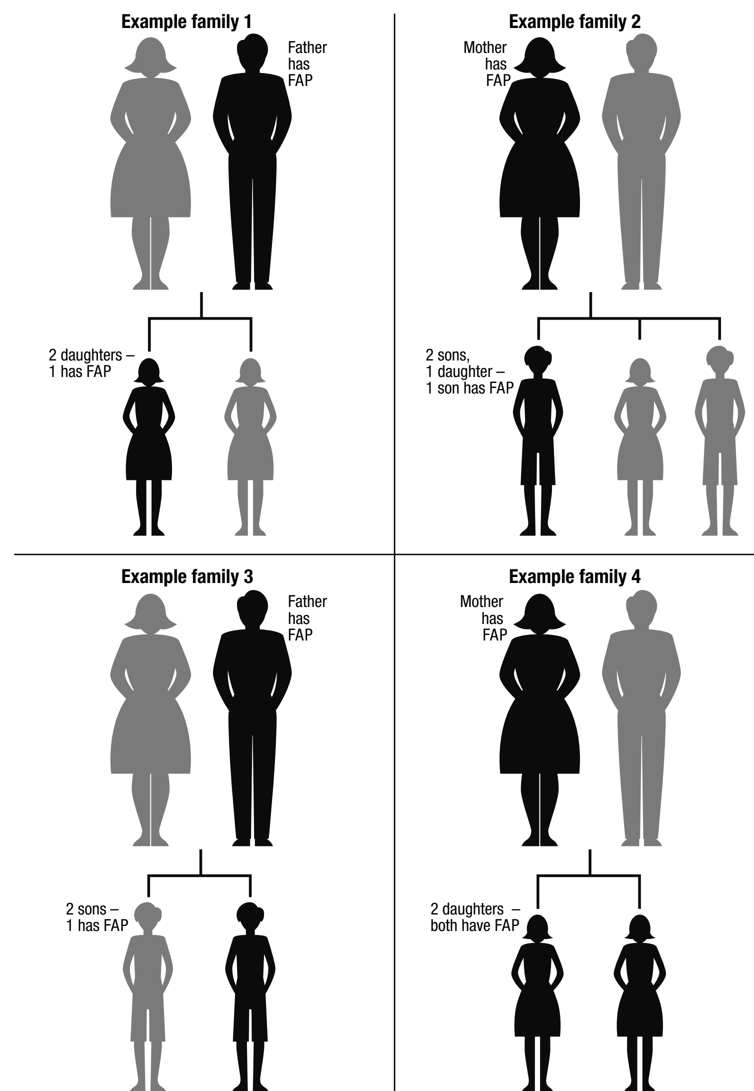
If I have FAP and two children, will one get the changed FAP gene and the other miss out?

One child might get it, or both, or neither. Each of your children has a 50:50 chance of inheriting your changed FAP gene. It's a bit like tossing a coin; each time, there's an equal chance of getting a head or a tail. In the same way, each time you have a baby, there's an equal chance of passing on the changed FAP gene or not.

One of my parents had FAP, but I don't. Are my children at risk?

It depends. To find out if you have the changed FAP **gene**, you would need to have a gene test (see page 13). If you are more than 55 years old and you haven't yet developed bowel polyps, it is unlikely that you have inherited the changed FAP gene from one of your parents (but only a gene test can confirm this). The changed FAP gene cannot skip a generation, so your children are not at risk if you don't have it.

Here are four possible FAP families, showing inheritance of the changed FAP gene. Many other family patterns are possible.



Remember: every child has a 50:50 chance of inheriting the FAP gene from a parent who has the FAP gene.

I have FAP, but nobody else in my family has had it. How can this happen?

About one in every four or five people with FAP has no known family history of the condition.

There are two possible explanations:

- One of your parents carried a FAP **gene** change in the egg or sperm cell which gave rise to you. Another possibility is that there was a gene change in the fertilised egg itself. In either of these situations, your parents would not be affected by FAP and you would have no family history of the disease. This type of gene change is called a new **mutation**.
- One of your parents had FAP, but it was never recognised. For example, they may have died from some other cause before bowel cancer appeared.

If it's not clear how you inherited your FAP, doctors usually err on the side of safety and suggest that any members of your family who might be at risk are tested. This includes your parents, brothers and sisters and your children.

Bowel polyps

Polyps are small growths, often on stalks like a mushroom. They vary in size from a tiny pinhead to two centimetres diameter or more. **Bowel** polyps are the most common sign of FAP: hundreds can develop, mainly in the large bowel.

Most people with FAP develop bowel polyps during their teens. The polyps may start at any age, but rarely before the age of ten.

Bowel cancer

Unless **bowel polyps** are removed, some will develop into bowel cancer. This usually happens in a person's late twenties, thirties or forties. In about six out of ten cases the cancer will have spread beyond the bowel by the time it is detected and will be incurable.

*** If people delay having check-ups until symptoms develop, then it is likely that some polyps will have already developed into cancer.**

With check-ups for FAP, the picture is very different. The aim of check-ups is to detect **polyposis** at an early stage, well before symptoms develop and then to arrange surgery to prevent bowel cancer (see pages 23–30). See your doctor regularly for a check-up.

*** The Cancer Council has a booklet on bowel cancer. Telephone 13 11 20 or visit www.cancervic.org.au**

Other signs of FAP

The other main signs of FAP are 'freckles' at the back of the eyes and **polyps** in other parts of the digestive system.

'Freckles' inside the eye

Many people with FAP develop pigmented patches (like freckles) inside their eyeball, on the **retina**. These are sometimes called **CHRPE** (pronounced 'chirpee').

These are not a problem: they don't affect vision, and they don't turn into cancer.

CHRPE can help the doctors to discover whether FAP is present, because the patches often appear at a young age, before **polyps** develop (though not everyone with FAP has CHRPE). To look for them, an eye doctor will put drops in your eye, then look into the eye. However, **gene** testing has made such eye examinations largely unnecessary.

Polyps in other parts of the digestive system

Polyps often develop in other parts of the digestive system.

Regular checks for polyps in the **duodenum** (duodenal polyps) are now widely done using an instrument called a duodenoscope.

Duodenal polyps can develop into cancer, but the risk is much lower and occurs later than for polyps in the large **bowel**. If you have indigestion, pain or weight loss you can't explain, if you are bleeding from the bowel, or if you become jaundiced, tell your doctor about it.

If the polyps turn into cancer, the treatment is surgery.

Research is still being done to decide whether, and how, duodenal polyps should be treated if they are not cancerous. New tests can find early-to-moderate stages of duodenal polyposis. Surgery is recommended for advanced stages of **duodenal polyposis**, even without cancer being present.

Not all polyps are due to having FAP. Polyps in the nose, uterus, (womb) or on the skin are quite common. These are nothing to do with FAP. If you are concerned about them, ask your doctor.

Gardner's syndrome

Gardner's syndrome is caused by the same **gene** as FAP. People with FAP often have one or more features of Gardner's syndrome. It varies from person to person, even within a family: one person might have plain FAP, another may have some signs of Gardner's syndrome.

People with Gardner's syndrome usually have small cysts and benign growths (**benign** means noncancerous) in various parts of the body. Often these occur in bones, particularly the jaw. In childhood, small lumps may appear below the skin, often on the face or scalp. They are not a problem, and don't usually need treatment.

Occasionally some teeth may be missing or impacted, or the person may have extra teeth.

A few people develop **desmoid tumours**. These are fibrous lumps, usually inside the abdomen. They are often slow growing, and may not produce symptoms for many years. They can become a problem as they get larger and affect blood vessels. They are not cancers, and do not spread to other parts of the body.

Attenuated FAP

Some families are thought to have a milder (or attenuated) form of FAP. These families develop fewer **bowel polyps**. Cancers occur, but later in life.

Management is much the same although **colonoscopy**, not just **sigmoidoscopy**, is required for check-ups as the **polyps** occur mainly on the right side of the bowel, out of reach of the sigmoidoscope. (For an explanation of sigmoidoscopy, see pages 20–21.)

Research into FAP

As more people with FAP are having surgery to prevent **bowel** cancer, we are learning more about the other symptoms of FAP and how best to manage them. Most of these symptoms appear many years after the bowel polyps.

Two other research areas are particularly interesting: drug treatment and genetic testing.

Drug treatment

We know that some drug treatments can reduce the size and number of **polyps** in the **bowel**, but we don't yet know how much they can help to prevent cancer. Research on this is continuing and your doctor is the best person to tell you about treatments that may help you.

Genetic testing

Knowledge in this area is growing rapidly. From the exact nature of the change in your FAP **gene**, doctors may in the future be able to tell more about how your particular FAP will develop – exactly where **polyps** will develop, and what other symptoms you might get. It is thought that other, as yet undiscovered, genes may modify the effect of the FAP

gene. This would explain differences in the pattern of polyp and cancer development between individuals and families.

The results of such research will help in deciding on the best treatment for each person.

In the meantime, you should always seek your doctor's advice about your own management, rather than waiting for alternatives.

Genetic testing for FAP

The specific change (mutation) in the FAP gene can differ from one FAP family to another, so the laboratory scientists need to find out what the change in the FAP gene in your family 'looks' like. The best way to do this is to take a blood sample from someone in the family who definitely has FAP, to search for and identify the **gene** change. This 'searching' is difficult to do, can take some time, and is not always successful.

Once the genetic change in a family (known as the family specific mutation) has been found, anyone else in the family who is at risk of FAP can have the gene test. This involves genetic counselling before having the test and is usually not done before the age of twelve.

This test result can tell you with certainty whether you have the changed FAP gene. People without the change in the gene will not develop FAP.

The test result

Negative: there is no change in the FAP gene

If the test shows that you do not have your family's FAP gene change (this is called a negative or normal result) you can be sure that you will not develop FAP. You cannot pass the changed gene on to your children as you do not have a changed copy of the gene to pass on. No further action is necessary.

A negative result means that you will not develop FAP but it does not mean that you will never get **bowel** cancer. Like everyone else in the population, you could still get bowel cancer. This affects about one person in twenty at some time, usually in later life.

Positive: the FAP gene is changed

If the test finds the FAP **gene** is changed – a 'positive' result – you will eventually develop polyps. It is vital that you have regular check-ups with your doctor to watch out for these and have treatment before bowel cancer develops.

Where is the gene test done?

Gene testing in Australia is done through special genetic clinics. These clinics make sure that people having genetic testing receive all the information and support they may need.

In Victoria, genetic counselling and genetic testing are available through the Victorian Family Cancer Genetics Service. You will need a referral from your doctor to attend one of these genetic clinics.

*** The Cancer Council Helpline can provide advice about contacting a family cancer clinic. Telephone 13 11 20.**

Cost and waiting for results

There is no charge for genetic testing.

Once you decide to be tested, you will probably want the result as soon as possible. Genetic testing is complex and specialised laboratory techniques are required.

This means that identifying the family gene may take many months. Ask the person doing your test how long it might take for your test. Once the gene change is identified, testing at risk family members is easier and quicker. In a small number of families, the gene change cannot be identified.

Talking it over first

Before you have the test, staff at the genetic clinic will talk it over with you. They will discuss what the test and its results might mean to you and to your family.

A *negative* result can be an enormous relief – but sometimes it can also be difficult, particularly if you know that a loved parent or brother or sister has the **gene** for FAP while you don't.

A *positive* result – finding out that you have the changed FAP gene – may be distressing. It can also be a relief, because it removes uncertainty. But you may feel angry, fearful and depressed. There will be many questions, such as, Will I get cancer? How will I cope with surgery? What about my children? What will friends think?

The genetic counsellor at the clinic can explain what to expect, and help you to sort through these thoughts and feelings.

I'm not sure if I want to know ...

This reaction is quite normal: the prospect of knowing for certain can be frightening. Genetic testing has many benefits, but it is not the best choice for everyone.

There are a few points to consider.

- If the test shows that you don't have the changed FAP **gene**, you know that you will not develop FAP.
- If the test shows that you have the changed FAP gene, remember that FAP can be treated successfully. It is important, however, to start treatment well before bowel cancer appears. With treatment, you have the chance of living a long and healthy life.
- If you have children, they have the right to know about FAP and its treatment, and about whether they are at risk or not.
- If you are the first to be tested in your family, your test result (with your consent) can be made available to all members of your family.

This is necessary for them to be tested for your family-specific change.

Talking with your doctor or a genetic counsellor who listens and answers your questions may help you decide if genetic testing is appropriate for you. It may also help to talk with another professional counsellor, or with someone else you trust and respect – family or friends, or your religious or spiritual adviser.

* **Professional counsellors may be available through your local hospital or community health centre. The Cancer Council Helpline can also put you in touch with counselling services. Telephone 13 11 20.**

Privacy matters

Genetics clinics ask a lot of questions about you and other members of your family. All your information is confidential. Your consent is needed before information about you can be passed on to any other person or organisation. Information about other family members will not be shared with you without their consent.

Who will know my test result?

With your permission, your test result will be sent to the doctor who referred you to the genetic clinic. If you have joined the Victorian Family Cancer Register, your test result will also be sent to the Register, as it could help in the future medical care of you and your family (see page 31 for information about the Victorian Family Cancer Register).

Two things are necessary in order to give correct advice to other members of your family:

- knowing about the FAP **gene** change in your family – what your family FAP gene 'looks' like
- knowing who in the family has FAP, so it can be shown whether or not others are at risk.

All information kept at the register is strictly confidential – it is never given to outside enquirers.

Insurance policies and genetic testing

If you are planning to have genetic testing, it is wise to discuss personal insurance policies before testing. This includes life insurance, disability (or income protection) insurance and income protection. Note that health insurance premiums are not affected by genetic testing or knowledge of genetic conditions. (Health insurance is community rated, meaning it uses the average per person premiums for everyone).

Your family history of FAP and any results of genetic testing may influence the cost and availability of life, disability and income protection insurance to you, but not private health insurance. Application forms for personal insurance will usually ask you to report any family history of inherited conditions and whether or not you have had genetic testing. They will also ask about your results.

Australian insurance companies have agreed that they will not ask applicants to have gene testing purely for insurance company purposes. Genetic testing clinics and the Victorian Family Cancer Register *will not* make your gene test result available to an insurance company without your written consent.

Genetic clinics can provide you with further information about the implications for insurance.

Can all FAP families have the genetic test?

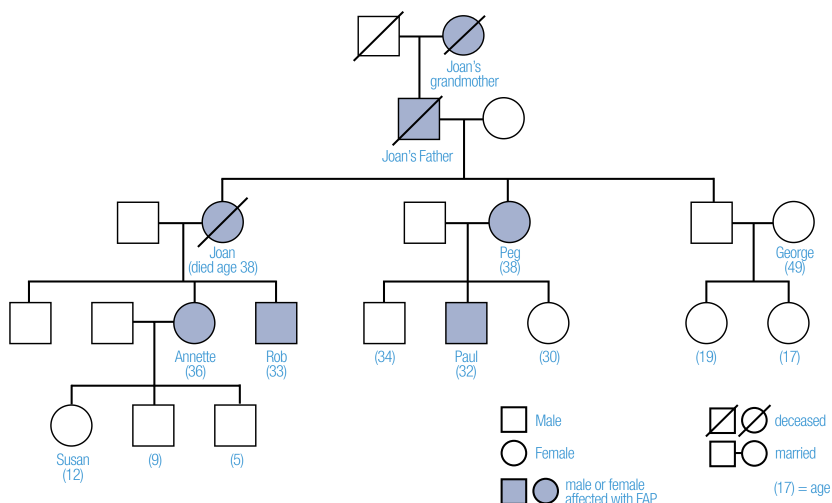
Sometimes, in a family with FAP, there is no one available who definitely has FAP: they may have died or be impossible to contact. This means the laboratory cannot get a blood sample from someone with FAP to identify the family gene, and so others in the family cannot be tested.

In a very small number of families, even though a blood sample is available, the laboratory cannot find the gene changes known to cause FAP. This means that others in the family cannot be tested.

If you are at risk of FAP, you will need to have regular **bowel** examinations, just as was done in all FAP families before the genetic test was available.

Annette's story

Annette was fourteen when her mother Joan, aged 36, was found to have FAP and **bowel** cancer. She was treated for the cancer, but lived only another two years.



Checking back, Joan and her doctor realised that Joan's father and grandmother had died young of bowel cancer. It was likely that this was an inherited condition. So Annette and her brothers had their bowels examined. She and a younger brother, Rob, were found to have polyps. Annette's family tree or 'pedigree' is shown above.

Symbols for family members with FAP are shaded. This is the standard method for drawing family pedigrees used by FAP doctors and genetic counsellors.

At the age of sixteen, Annette had surgery to remove her bowel. She has had yearly check-ups with **sigmoidoscopy** ever since and remains fit and healthy. Rob had the same operation three years later, and he too remains well.

Joan and her parents and grandparents did not know about FAP and their risk of inheriting it, and so were never checked for it. It was only Joan's bowel cancer that brought it to light.

The younger generations are luckier. When Joan was diagnosed, her sister Peg and Peg's children were also checked. Peg and her son Paul (Annette's cousin) were found to have FAP. Both have had bowel surgery. So far, however, Peg's other two children (both now in their thirties) have not developed polyps: they probably don't have FAP. (They have chosen surveillance, and don't want genetic testing yet.)

Joan's younger brother, George, did not want to know about it and never went for check-ups with the doctor. He has been lucky: he is now nearly fifty years of age and remains healthy. He probably doesn't have FAP, but it is possible that he does, and that **polyps** will develop into bowel cancer. His two teenage daughters still don't know whether they are at risk.

Annette is now married with three children. Only recently, she had a blood sample taken to identify the family FAP **gene**. She has discussed FAP with her children. Her eldest daughter, Susan, now twelve, has decided to have the genetic test between her fourteenth and fifteenth birthdays.

Once the gene change that causes FAP has been identified for this family, genetic testing can also be offered to Peg's other children, and even to Peg's brother, George. If George does not have the changed FAP gene then he and his daughters will not develop **polyposis**.

Regular check-ups and seeking support

If you are in one of the following groups, you need to see your doctor regularly to check for polyps in the bowel.

If you have the changed FAP gene

You should see your doctor and have your bowel looked at every year to check for polyps. When **polyps** appear, you will need to discuss surgery with your doctor.

If you are at risk and have not had the gene test

Your **bowel** should be checked every one to two years until you are thirty to thirty-five years old. After this, the risk becomes less and a check-up every three years is enough.

If you have had surgery for FAP

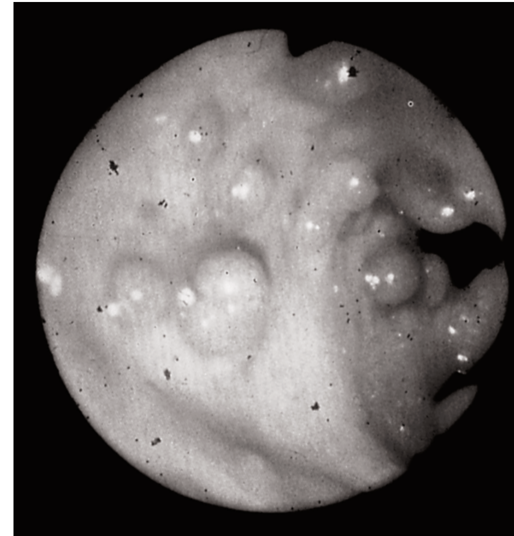
Most people will still need regular check-ups after surgery for FAP. See 'Surgery to prevent bowel cancer' (page 23–30), and 'Other signs of FAP' (page 9).

What is involved in check-ups?

A doctor (usually a gastroenterologist) will look into your bowel using a sigmoidoscope. This is a small, tube-like telescope with a light at its end. It is inserted gently through the **anus** into the first twenty-five or sixty centimetres of bowel (the distance depends on the type of sigmoidoscope used). This is called **sigmoidoscopy**. It can be slightly uncomfortable, but it should not hurt. It takes only a few minutes.

Before the test you may need a bowel **enema**: fluid is passed into the bowel to cause a bowel action. Clearing out the bowel helps the doctor to see any **polyps** clearly.

Polyps seen through a sigmoidoscope



If the doctor sees polyps during the examination, a small sample (called a **biopsy**) will be taken. This is not painful and cannot usually be felt. The sample is sent to the laboratory for analysis.

Your **duodenum** also needs checking with a side viewing gastroscope. This should happen every five years. If polyps are found, then it should happen every two years. If polyps are dense, surgery may be considered and/or endoscopic resections (removing polyps with the aid of an endoscope).

Cancer Council Helpline

The Cancer Council Helpline is a free, confidential service where you can talk about your concerns and needs with a cancer nurse. They can send you information and put you in touch with support services in your area. Telephone 13 11 20.

Multilingual Cancer Information Line

The Multilingual Cancer Information Line is a free and confidential service of the Cancer Council. You can call and speak to a cancer nurse with the help of an interpreter. It is for people with cancer, and people who are close to them. People who speak any language can use the service. See the inside back cover for details.

Surgery to prevent bowel cancer

The treatment for FAP is to remove all or most of the large bowel (the colon and sometimes the rectum) after bowel polyps start to appear (see diagram page 4).

The aim is to prevent cancer from developing and, at the same time, to keep your digestive system working as normally as possible. Losing your large bowel does not affect how your body absorbs food.

Talking to your doctors

Getting used to the idea of surgery is not always easy. You should talk to your doctor about what is involved.

You have the right to ask your doctor anything you want to know. If you don't understand what the doctor says, ask them to explain it in a different way – doctors often use technical words without realising it. It can help to write down your questions first, and take notes of what the doctor says. You may choose to ask either your GP or your specialist physician or surgeon.

You may find it helpful to talk with someone who has had the same operation. This can be arranged through your surgeon, the FAP coordinator or through the Cancer Council Helpline.

In FAP families, there is often someone else in the family who has had the operation and it may be useful to talk to them.

You may also wish to talk about the social and emotional impact of surgery with a social worker or professional counsellor, before and/or after the operation. This type of help is often available through your hospital. The FAP coordinator (9635 5176) or the Cancer Council Helpline (13 11 20) can also put you in touch with a counsellor.

Delaying surgery

There are strong arguments against putting off surgery until **bowel** cancer develops.

- Even with regular examination, it is very difficult to find a small cancer among hundreds of **polyps** that can develop in the **colon** in FAP. Chances are your cancer would not be found until it was too late to be cured.
- You would need to have a **colonoscopy** at least every year. This is time consuming and may be uncomfortable.

However, there is a trend to delay surgery until the early to mid-twenties, if the polyps are numerous but small.

If there is a strong history of **desmoid** disease in the family, surgery in the colon might best be avoided altogether, given the risks of surgery for these people. Drugs may be an effective alternative treatment.

Treatment options

Three different operations may be done for FAP. Each has advantages and disadvantages. Deciding on the best option is not always straightforward. It will depend largely on your age and the stage of your FAP.

You should talk to your doctor about the best treatment for you.

Total colectomy and ileorectal anastomosis (colectomy and IRA)

This is the most common operation. The surgeon removes all the **colon** ('total **colectomy**'), and then the end of the **ileum** is joined up to the **rectum** ('**ileorectal anastomosis**').

After recovering from surgery, most people find they have two to three **bowel** actions a day.

The *advantage* of the operation is that you keep your rectum and **anus**. You continue to go to the toilet to pass bowel motions in the usual way.

The *disadvantage* is that there is still a risk of **polyps** and cancer in the rectum. This means regular appointments with the doctor for the rest of your life to examine the rectum and remove any small polyps before they become a problem.

Studies suggest that drug treatment may help to slow the growth of polyps in the rectum after the operation. However, the benefits and risks of this treatment are not yet known for certain.

Restorative proctocolectomy

This is the same as a colectomy and IRA (see above), except that the surgeon also removes most of the **rectum** and all of its lining.

A 'new' rectum (called a pouch) is made from the end of the **small bowel**, and this is joined up to the **anus**. You may have an **ileostomy** for a short time (at least two months) until the pouch surgery heals.

The *advantage* is that, with the lining of the rectum gone, there is no danger of **polyps** or cancer forming there. After the temporary ileostomy has gone, you will go to the toilet to pass bowel motions in the usual way.

The *disadvantage* is that many people have five to seven loose bowel actions a day. The stools will be the consistency of porridge. It should be no problem to hold on for a while after feeling the urge to go. However some people need to go at night and may feel more confident if they wear a small pad in case any leakage occurs. Other problems, such as post-operative infections and **adhesions**, are also more common. Further polyps commonly develop in the pouch after about seven years.

Proctocolectomy and ileostomy

This is rarely done now. It may be done if there is already a cancer low in the **rectum**, or if the person is **incontinent** after one of the other two operations.

In this operation, the surgeon removes all of the **colon**, rectum and **anus**. The end of the **small bowel** (the **ileum**) is then joined to the wall of the abdomen, opening to the outside to allow bowel motions (**faeces**) to pass out. The hole itself is called the **stoma** (the word simply means an opening). If you have this operation, you will need to wear a special bag over the stoma to collect the bowel motions. **Ileostomy** is discussed later in this booklet.

After the operation

With early detection and treatment there is every chance that you will continue to live a full and healthy life.

The main difference for most people is more frequent **bowel** motions. The operation does not usually affect fitness, social or sexual activities or the ability to have children.

If you do have problems – for instance, in managing diarrhoea – there are many approaches that can help. You should talk these problems over with your doctor.

How long does it all take?

Most people are in hospital for about ten days, although it can be up to about three weeks, depending on the operation.

If you have a pouch, you may need a few extra days in hospital for the new piece of **bowel** to settle down.

Check with your surgeon how long you should allow before returning to your usual level of activity. This will depend on the type of surgery and the family, social, work and sporting commitments that you have. People who are working are usually back at work after about six weeks. Students often find that the long vacation between secondary school and tertiary education is a good time to have the operation.

After a few months, your bowel gets used to being shorter and adapts. In the meantime, there are various medicines that can help with diarrhoea. Medicines to control diarrhoea can be particularly useful if you are under stress or need to go to a special event.

Regular check-ups

After the operation, you may still need regular check-ups with your doctor.

If you have had a total **colectomy** and IRA (see pages 24–25), you are still at risk of developing **polyps** and cancer in the **rectum**. With regular check-ups, your doctor can remove any polyps that develop.

If you have had one of the other operations, there is no chance of developing cancer in the **large bowel**. There is still, however, a risk that you might develop polyps or other features of FAP somewhere else in your body, including in the pouch. Regular checks for these, especially for polyps in the upper part of the **small bowel** and the pouch, are advised. See the section titled 'Other signs of FAP'.

Diet

After **bowel** surgery, you may need to make a few changes in your diet, particularly if you have an **ileostomy**.

Foods to be careful with include 'windy' foods such as cabbage, onion and Brussels sprouts; 'rough fibre' foods such as seeds, nuts and heavy whole grains; 'pithy' foods such as oranges; and rich, fatty foods.

Fibre may make an ileostomy act more often than usual. But it is important to include food with fibre: try smoother fibres such as bran, bran flakes and fruit.

It is also important to drink six to eight cups of water a day.

Different people may react to different foods. Most people have a few months of trial and error to find out what foods suit them best. Try introducing foods one at a time.

Some food reactions may be temporary; after a while you may find the same foods are not a problem. It may take up to twelve months until your bowel settles down completely.

If you need information about diet after surgery, your doctor can discuss this with you or refer you to a dietitian.

*** The Cancer Council's booklet *Nutrition and Exercise* contains useful information. Telephone 13 11 20 or visit www.cancervic.org.au**

Living with an ileostomy

Learning to look after an **ileostomy** takes time, patience and practice.

But you should be able to return to almost everything you did before the operation: work, most sports or exercise (including swimming), social life, holidays and clothes you enjoy wearing.

A specialist nurse called a stomal therapy nurse will talk with you before the operation if you wish. The stomal therapy nurse will help the surgeon to decide exactly where the **stoma** should be, so that your bag will stay in place whether you are sitting, standing or moving about.

You may find it helpful, either before or after the operation, to talk over the practical and personal aspects with someone who has already learned to live with an ileostomy. Your doctor or stomal therapy nurse may be able to arrange this.

The new stoma will be slightly swollen. It can take several weeks to settle down to its normal size – a small, slightly raised, neat, pink opening. Before you leave hospital, the stomal therapy nurse will teach you how to fit and change the ileostomy bag and look after the stoma, and will help you to regain your confidence.

Once you are at home, you can phone the stomal therapy nurse about any problems, whether they arise days, weeks or even years later. The Royal District Nursing Service also has stomal therapy nurses who can help you after you leave hospital.

In some areas, it is possible to buy custom-made garments that hold an ileostomy bag neatly and firmly, for instance, attractive underwear for women and bathers and underpants for men. Ask your stomal therapy nurse, or contact the Cancer Council Helpline for details. Telephone 13 11 20

Relationships, sex and bowel surgery

On the physical side, surgery for FAP, even having an **ileostomy**, should not affect your ability to enjoy sex. For more practical information and support on sexual activity with an ileostomy, contact your stomal nurse or the Cancer Council Helpline.

For some people, surgery can change the way they feel about themselves and their bodies. Or they may be very concerned about how their partner will cope. It can affect their whole relationship. As many people diagnosed with FAP have surgery when they are still quite young, this may impact on their developing self-esteem, body image and feelings of attractiveness.

A leaky **bowel** or an ileostomy can cause people to feel different or self-conscious.

*** The Cancer Council has information on sexuality and cancer. Telephone 13 11 20 or visit www.cancervic.org.au**

If you are in a relationship, talk things over with your partner: this is one of the most important things you can do. Good, open communication goes a long way to sorting out any problems that exist. It will also reassure both of you of your affection and need for each other.

If you are in a new relationship, telling others about bowel problems can be difficult. When do you tell? It is probably best to know people well before you tell them. A bowel problem is a much smaller issue for someone who already knows and cares for you. There may be particular challenges for you if you are just learning how to negotiate

relationships. Remember that you are in control of the situation. You can choose who to tell, and when to tell them.

It can help considerably to discuss these problems with someone outside your relationship. Another person can see your situation differently and may suggest new ways that you might manage it. This person might be your doctor, the stomal therapist, or someone at home or in your community who can help. There are also excellent and experienced sexual counsellors available, who may be social workers, psychologists or psychiatrists.

Call the Cancer Council Helpline on 13 11 20 for details.

Pregnancy and childbirth

Pregnancy and childbirth may pose problems for women after **bowel** surgery.

Women who have had restorative proctocolectomy may be less likely to conceive. This can be important if you need to have surgery before completing your family. Conception rates are much less affected with total **colectomy** and **ileorectal anastomosis**.

Some time should be allowed between having surgery and becoming pregnant and there may be some types of bowel surgery where a caesarean delivery would be advisable. These matters should be discussed with your doctor and obstetrician, as each situation will be different.

The Victorian Family Cancer Register

People with FAP may join the Victorian Family Cancer Register. This includes the FAP Register. The register helps those with FAP and other families with an increased risk of developing cancer because of an inherited change in a **gene**.

The Victorian Family Cancer Register is kept at The Cancer Council Victoria. The Registration and Surveillance Coordinator looks after registered FAP families.

The Victorian Family Cancer Register can:

- send you reminder letters when your **bowel** examinations are due
- send you a regular newsletter
- send you updates of this FAP booklet
- provide information on FAP-its diagnosis and treatment and the FAP genetic test
- be a point of contact for people and families with FAP
- contribute to research which may help you and your family in the future
- work with doctors, family cancer centres and other FAP Registers looking after you and your family. If people know they are at risk of FAP, medical advice and care is available.

What information will be kept if I join?

With your consent, the following information will be kept:

- your name and date of birth
- your position in the family
- your contact details.

The following information will also be kept, if it applies, and you consent:

- FAP **gene** test results
- whether you have had cancer and at what age
- details of cancer treatments and their results
- cancer screening tests and results.

Protection of personal information

All written information that identifies you is stored in an area with access restricted to Victorian Family Cancer Register staff. All electronic records that identify you are stored on a password protected computer system.

You have access to your own information but no information about you is given to your family members. If you have agreed, the FAP coordinator may give information to doctors, genetic counsellors or family cancer centres involved in the care of your family members.

Joining the Victorian Family Cancer Register

The FAP coordinator at the Cancer Council (9635 5176) can tell you more about the Victorian Family Cancer Register and how to join. The coordinator works with doctors, hospitals, family cancer centres, and patients and their families in Victoria.

Caring for someone with cancer

A carer is anyone, whether family or friend, who is helping to look after someone with cancer. Caring for someone with cancer can be very stressful, particularly when it is someone you care about very much. The person with cancer may be experiencing distressing emotions about their cancer **diagnosis**, side effects from treatment and mood changes from the effects of drugs.

Look after yourself during this time. Give yourself some time out, enjoy a cup of coffee with a friend, and share your worries and concerns with some one not involved. Make a list of 10 things you like to do and make sure that you do one enjoyable thing each day.

You may have to make many decisions. You will probably have to attend many appointments with doctors, support services and hospitals. Many people have found it helpful to take with them another member of the family or a close friend. It also helps to write down questions before you go, and to take notes during the appointment.

Cancer support groups are usually open to patients and carers. A support group can offer the chance to share experiences and ways of coping.

There is a range of support services such as home help, meals on wheels and visiting nurses that can help you cope with treatment at home. These are provided by local councils, the Royal District Nursing Service and the palliative care services.

Call the Cancer Council Helpline on 13 11 20 to:

- be linked with another carer by telephone
- speak with a cancer nurse for further support and information
- be sent a carer's kit so that you can find out about financial assistance and other resources.

* **The Cancer Council has a booklet for people caring for someone who has cancer. Telephone 13 11 20 or visit www.cancervic.org.au**

Questions to ask your doctor

If you have FAP you may find this list helpful when thinking about the questions you want to ask your doctors about your condition, bowel cancer and its treatment.

Current status

1. What is my risk of developing bowel cancer?
2. Is there a risk of cancer in other parts of my body?
3. Do I already have polyps? What kind are they?
4. Can I reduce my risk of developing bowel cancer?
5. Is diet important?
6. Are there any other issues I need to be aware of?
7. Can you explain to me the result of my gene test?

Surgery

1. Will I need surgery? If so, when?
2. What are the risks of surgery?
3. Will I have any lasting side effects?
4. Will I need a colostomy; if so, will the stoma be permanent?
5. How will I feel after the operation?
6. If I have pain, how will it be controlled?
7. How long will I be in hospital?
8. How will surgery affect my normal activities? Am I likely to have urinary problems? What about bowel problems such as diarrhoea or rectal bleeding?
9. Will treatment affect my sex life?
10. When can I get back to my normal activities?

Regular check-ups

1 Why do I need to have regular check-ups?
2 What check-ups should I have?
3 How often do I need them?
4 Can you remind me about my check-ups?
5 Where will I have the check-ups?
6 Are the tests painful?
7 How soon after the tests will I learn the results?
8 If polyps are found, what happens
9 If I have check-ups, can I still get cancer?
10 If I develop cancer, who will talk to me about the next steps?
11 Is there anything else I should look out for?

Family

1 Are my children at risk of FAP?
2 Do they need a gene test?
3 Should I bring them to see you?
4 What treatment will you advise for them?
5 When would it start?
6 I want to have a baby, but I don't want the baby to have FAP. What are my options?
7 Should I tell other family members about FAP? Can you help me with this?
8 I don't speak to my sister anymore – can someone else tell her about FAP?
9 Are my brothers or sisters at risk of bowel cancer?
10 Which of my family members should see you about treatment?

If there are answers you do not understand, feel comfortable to say, 'Can you explain that again?' or 'I am not sure what you mean by ...'

Glossary: what does that word mean?

Most of the words listed here are used in this booklet; others are words you are likely to hear used by doctors and other health professionals who will be working with you.

adenoma A benign tumour (not a cancer) that may turn into a cancer if not treated.

adhesions Bands of fibrous tissue growing abnormally in parts of the body that may cause pain, nausea, bloating or an inability to pass gas, and constipation.

adenomatosis polyposis coli (APC) A gene that is mutated in people with familial adenomatous polyposis (FAP).

anastomosis The joining together of two tubes, such as two cut ends of the bowel.

anus The opening of the bowel through which bowel motions are passed.

benign Not a cancer.

biopsy The removal of a sample of tissue from the body, for examination under a microscope, to assist diagnosis of a disease.

bowel The long, tube-shaped organ that extends from the stomach to the anus. It absorbs food and water and passes the waste products of digestion as faeces.

CHRPE Pronounced 'chirpee', these are harmless pigmented patches inside the eyeball on the retina. They are common in FAP. CHRPE stands for 'congenital hypertrophy of the retinal pigment epithelium'.

colectomy The surgical removal of the colon.

colon The part of the large bowel between the end of the small bowel (the ileum) and the rectum. It is about 1.5 metres long.

colonoscopy Examination of the large bowel using a colonoscope. This is a thin, flexible tube with a light at the end. It is passed through the anus and gently moved around so that through it, the doctor can see the full length of the large bowel.

desmoids Fibrous lumps or tumours occurring disproportionately, though uncommonly, in FAP.

dominant inheritance Only one altered gene is required (from mother or father) for disease to develop.

duodenoscopy A flexible tube examination of the duodenum, usually with a side viewing endoscope.

duodenum The first thirty centimetres of the small bowel. The stomach empties into the duodenum.

endoscope Any instrument allowing visualisation of a hollow tube structure (such as the bowel or stomach).

enema Fluid is passed into the bowel via the anus (back passage), in order to cause a bowel action.

faeces Bowel motions; waste matter passed from the gut via the anus (back passage).

gastroenterologist Doctor who specialises in treating diseases of the digestive tract.

genes The tiny factors that control the way the body's cells grow and behave. Each person has a set of many thousands of genes inherited from both parents. These genes are found in every cell of the body. The gene that is associated with FAP is called 'APC'.

ileorectal anastomosis The ileum and rectum are joined after the colon is removed.

ileostomy An opening through the abdominal wall through which the ileum is brought to replace the function of the anus. An ileostomy is performed after the surgical removal of the colon and rectum.

ileum The lower half of the small bowel, which joins up with the colon.

incontinent Involuntarily passing faeces or urine.

intestines See bowel.

jejunum The part of the small bowel below the duodenum and leading into the ileum.

large bowel The colon and rectum.

malignant Cancerous. A malignant tumour is a cancer. It tends to spread, and eventually causes death if it is not treated.

mutation A change in a gene, causing it to show a new characteristic.

polyp An abnormal growth or adenoma, often on a stalk.

polyposis The condition of having large numbers of polyps in the large bowel.

prognosis An assessment of the course and likely outcome of a person's disease.

rectum The last twelve to fifteen centimetres of the large bowel, which opens to the outside at the anus. The faeces collect in the rectum before they are passed as a bowel motion.

retina The light-sensitive lining inside the eyeball.

sigmoid colon The last twenty to twenty-five centimetres of the colon, which leads into the rectum.

sigmoidoscopy Examination of the rectum and sigmoid colon using a sigmoidoscope. This is a narrow tube with a light at the end. It is inserted gently through the anus, and gives a view of the lining of the bowel.

small bowel The part of the bowel comprising the duodenum, jejunum and ileum.

stoma An artificial opening created in the body by surgery. For example, an ileostomy is a stoma made where a cut end of the ileum (or small bowel) is brought through the wall of the abdomen and opens to the outside.

tumour A new or abnormal growth of tissue on or in the body.

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