Chronic Myeloid Leukaemia

WHAT IS CML? Chronic myeloid leukaemia (CML) is a type of cancer that affects the blood and bone marrow. In CML the bone marrow produces too many white cells, called granulocytes. These cells, sometimes called blasts or leukaemic blasts, gradually crowd the bone marrow, interfering with normal blood cell production.[1]

CAUSES AND RISK FACTORS

There are no specific causes identified for the development of CML but the disease results from a genetic abnormality in the myeloid stem cells (called the Philadelphia (Ph) chromosome). There is no evidence that the genetic mutation is inherited.[2] Most patients present with no known risk factors. However, some risk factors identified for CML include:[2]

AGE: The risk of getting CML increases with age

GENDER: This disease is slightly more common in males than females, but it's not known why.

RADIATION EXPOSURE: Being exposed to high-dose radiation (such as being a survivor of an atomic bomb blast or nuclear reactor accident) increases the risk of getting CML

There are no other proven risk factors for CML. The risk of getting CML does not seem to be affected by smoking, diet, exposure to chemicals, or infections. CML does not run in families.

SYMPTOMS

CML is not always obvious in the early stages. Some patients live with the condition for months or years before diagnosis. Common symptoms include: [1,3]

- Frequent infections
- Weight loss
- Tiredness, breathlessness or looking pale due to a shortage of red blood cells (anaemia)
- Loss of appetite
- Excessive sweating
- Abnormal bruising or bleeding
- Swollen lymph glands
- Abdominal discomfort or indigestion due to an enlarged spleen

EPIDEMIOLOGY AND PROGNOSIS

Each year in Australia around 330 people are diagnosed with CML. Overall, it is a rare disease, accounting for only 0.03% of all cancers diagnosed.[1]

These days the prognosis for CML patients is very good – it is hoped and expected that most patients will be able to live a normal life and have a normal life span. They will probably require lifelong tablet medication.[1,4]

Treatment is likely to involve the use of a type of tyrosine kinase inhibitor (TKI) - which blocks the leukaemia-causing effects of a substance called a tyrosine kinase, forcing cell apoptosis, or cell death.[1]

A stem cell transplant may be an option for some younger patients, or patients who are intolerant or resistant to TKIs - providing them with a better chance of cure.[1]

TREATMENT

Speaking generally, CML treatment will vary in patients depending on disease phase, general health and patient age .

There are three phases of CML – chronic, accelerated and blast phases.

The **chronic phase** is when the leukaemia is most stable and still developing slowly.[1] It is estimated that around 9 in10 people (90%) are in chronic phase at diagnosis, when symptoms may be mild or vague.[3]

In the **accelerated phase** patients may have more obvious symptoms, and may be more tired than usual or have lost weight.

The spleen may be enlarged, sometimes causing an uncomfortable or painful feeling on the left side under the ribs.[3]

The blast phase is also called the acute phase, blast crisis or blast transformation. This is when the leukaemia transforms into an acute leukaemia (usually acute myeloid leukaemia).[3]

Symptoms are troubling and patients feel unwell.[3]

CML OUTLOOK

PROFESSOR TIM HUGHES, AUSTRALIAN LEUKAEMIA AUTHORITY

"CML is a disease that can strike at any age but is most commonly seen in those over 50. Most patients have no known risk factors when they first present.

CML represents about 30% of the leukaemias that we see in Australia.

At the moment there are about 3000 patients that are being treated with kinase inhibitors in Australia.

For a patient with CML, the scenario that was in front of them in the 1990's was extremely bleak. Half the patients died from their leukaemia within three years. We now have a situation where these kinase inhibitor therapies have transformed the outlook, so that most patients can expect to live a normal life span and have a normal quality of life.
We still have challenges, but we now have an effective treatment for almost every patient with CML."

** Professor Hughes commentary was provided in November 2015.

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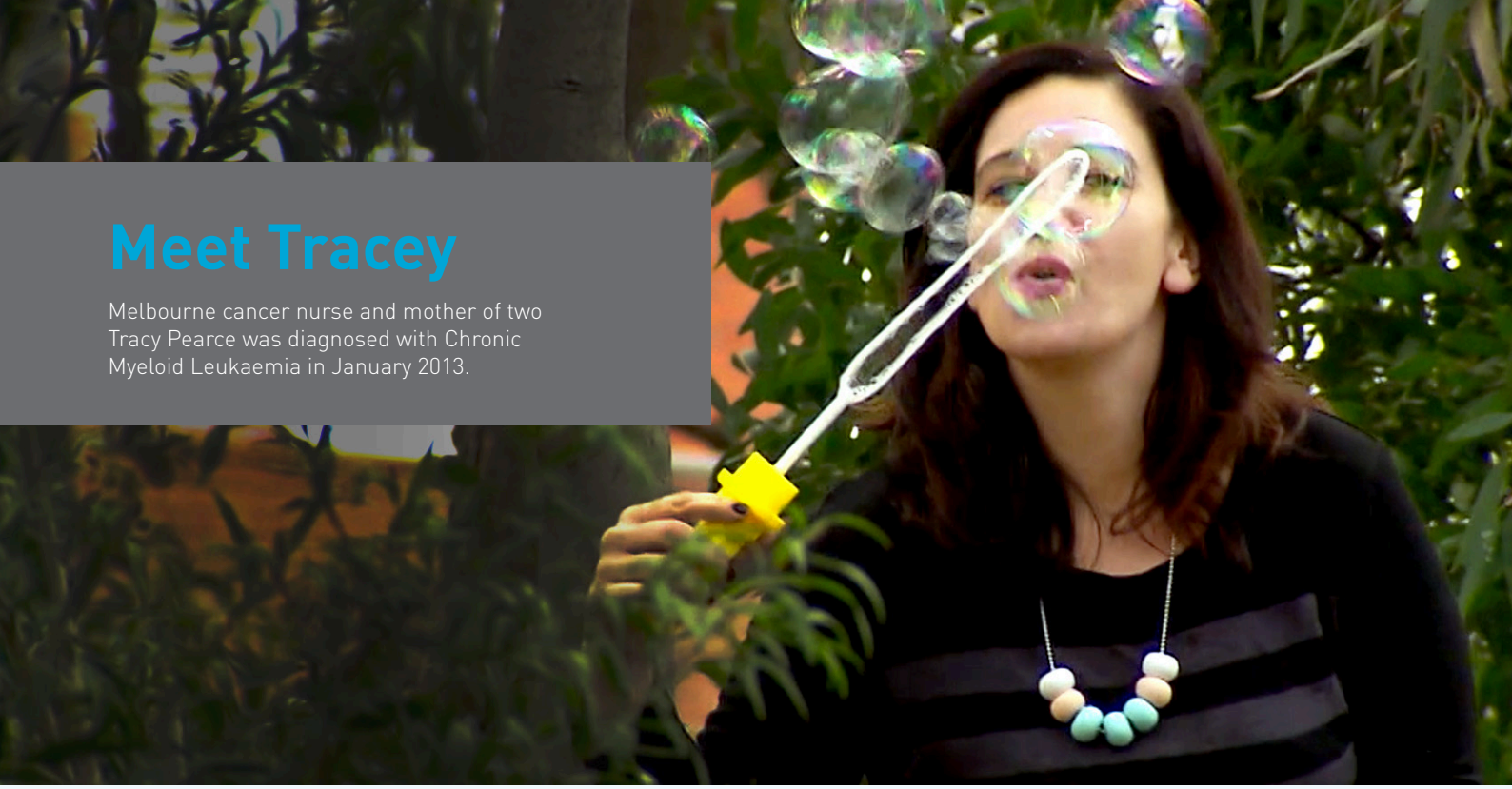

Meet Tracey

Melbourne cancer nurse and mother of two Tracy Pearce was diagnosed with Chronic Myeloid Leukaemia in January 2013.

"Leading up to it I was quite unwell for several months with intense headaches and some fatigue. Closer to when I was diagnosed I had some bruising and a chest infection I could not get rid of.

As a cancer nurse, I was quite aware of the symptoms, but I think when you are working with it every day and my young age, I wasn't expecting it to happen to me

I had two small children and I had just returned to work from maternity leave, so I just thought I was not coping very well, with having two young children and having sleepless nights and trying to get into a routine with my family.

My GP ordered a set of blood tests and I received a telephone call requesting that I go back to the clinic. I said I was still working and they impressed upon me it was more urgent than that. I got there and the GP told me the news that I had CML. I was devastated, numb, in disbelief. And I had an overwhelming sense of fear that I could die from CML. It was like the world had just stopped.

It wasn't until I knew more about CML and how positive the prognosis is, that I felt a bit better.

I began oral chemotherapy then I got into a randomised controlled trial for a newer drug.

I experienced significant side effects the first few months (like nausea and vomiting) but they did subside. And because I was part of a trial I was very well monitored.

I reached remission three to four months post treatment, which was really good. The initial prediction was remission in 12 to 18 months post treatment.

Following termination of the trial, I switched to an alternative drug and I remain in a functional remission.

My disease is well and truly at bay. To stay that way I need to be on therapy for the rest of my life.

I am in a really good place. I feel very fit and well and I am tolerating my daily treatment and I am working in a full time job.

It is a great place to be in. I am looking forward to a very long life with my husband, raising our children and sharing in their lives.

When I first started looking after patients well over a decade ago, these patients had a very poor prognosis. We have come so far in being able to manage this disease – it is really, really exciting."

* Tracey shared her story in February 2016.

REFERENCES

1. http://www.leukaemia.org.au; Last Accessed July 2016
2. http://www.cancer.org; Last Accessed July 2016
3. http://www.cancerresearchuk.org; Last Accessed July 2016

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