**Ehlers-Danlos syndrome accessible transcript:**

**Fraser**

**Visual**

**A man in his 50s, Dr Fraser Burling, Rheumatologist, sits on a dark grey striped couch. He has balding dark grey hair and wears a white shirt and tie, a black, grey and maroon argyle patterned V-neck jumper and a navy jacket. Pinned to the lapel of Fraser’s jacket is a looped campaign ribbon with black and white zebra stripes on it.**

**Audio**

My name is Fraser Burling. I'm a dual-trained physician and rheumatologist, and I work at the Rheumatology and Musculo-skeletal Clinic in Auckland. We had two patients come to the clinic with Ehlers-Danlos syndrome, one of which was Tracey. And treating them for their soft tissue injuries with prolotherapy, which is a process of strengthening damaged ligaments, resulted in significant improvements in those two.

**Visual**

**White text on a dark blue screen with a green border around it reads ‘Fraser explains the significance of the zebra symbol for Ehlers-Danlos syndrome (EDS) and the important message behind it.’ The green border shrinks and the text disappears. Fraser looks down, touching his jacket lapel.**

**Audio**

This ribbon represents the zebra, which is the mascot for Ehlers-Danlos syndrome. The first reason is if you hear galloping hooves, don't automatically think 'horses'. The second reason is the clinical expression for Ehlers-Danlos syndrome is as varied as the stripes of a zebra, and no two zebras have the same stripes.

**Visual**

**White text on a dark blue screen with a green border around it reads ‘Fraser explains the complex presentation and assessment of EDS and the role of collagen.’ The green border shrinks and the text disappears.**

**Audio**

Ehlers-Danlos syndrome is a collection of conditions of abnormal collagen synthesis,

whether it be the collagen molecules themselves or the molecules that help put collagen together. We currently have 30 known genes for Ehlers-Danlos syndrome. So Ehlers-Danlos syndrome, being about collagen synthesis — collagen holds everything together in your body, not just joints and bones, but it also holds together the soft tissues, such as your muscles, your blood vessels, your internal organs. And, in fact, pretty much everything in the body is held together by collagen. So when you have abnormalities within Ehlers-Danlos syndrome, it's not just the musculoskeletal abnormalities, such as being able to put your thumb through to the wrist or being able to bring this little finger back to the midway or stretching the arm back past 180 degrees, which is where you see with part of the Beighton's score.

**Visual**

**White text on a dark blue background with a green border around it in the lower third of the screen reads ‘The Beighton score is a popular screening technique for hypermobility. This is a nine-point scale and requires the performance of five manoeuvres.’ The green border shrinks and the text disappears.**

**Audio**

But it is also about the skin pulling back further, and in many people with the classical types, it is about people dropping their blood pressure on standing, because the vessels in their legs can't push the blood back up properly. It's about trouble with swallowing food for some people, with digesting food for some people, with pushing the food out through the back passage so that people can be constipated for up to six weeks at a time without a bowel motion. It really affects multiple different aspects of the body. We then look at multiple other issues that are involved, so I actually have a whole checklist page that I go through. So asking things like, 'Were you able to do party tricks when you were younger?' So these include the ability to do the prayer hands behind your back, the... Some people could put their foot over their head. Many people could do the splits without having to work towards doing that. People would often show off to their classmates in school about bending their fingers back or bending part of the finger.

So they'd be able to do, sort of, that motion with their finger without any effort at all.

Then there were those who would actually pull on their skin and pull it out. Then their history, such as whether they bruise easily or not, whether they feel faint or dizzy on standing. Some subtypes have a Marfanoid habitus, which means that they have extra-long fingers, extra-long toes, extra-long limbs compared to their height. When you're looking at the classical phenotypes of Ehlers-Danlos syndrome, it was, a long time ago, thought to be one in 75,000, then one in 150,000. Then it was brought down to one in 10,000. More recent literature has put it in one in 5000, and very modern literature is putting it more like one in 2500. And it may end up being closer to one in a thousand. We don't know yet. I think it's been more an issue of it being underdiagnosed, rather than not existent. The fact that we have multiple issues that are now recognised and multiple treatment options that are also recognised and are able to be applied to patients so that they no longer have to live in chronic pain, and many of them can be made pain-free. So, for example, I had one patient who took 10 minutes just to get out of a car and then two years later was able to climb a mountain — overseas. And she was able to do that because of the fact that we'd got her through the pathway with a multidisciplinary team approach and got her back to living a normal life again.

**Visual**

**White text on a dark blue screen with a green border around it reads ‘Fraser explains that people injure far easier with Ehlers-Danlos syndrome.’ The green border shrinks and the text disappears.**

**Audio**

Now, this isn't a one-off. Many of our patients come to us on the verge of suicide, because their life has been destroyed by injuries. Now, let me make it clear here. Ehlers-Danlos syndrome does not cause injury. Accidents cause injury. But people with Ehlers-Danlos syndrome are more easily injured when they have an accident and they're harder to heal. And so by the process of a multidisciplinary team approach, working together, we've got many people back to living a normal life again, or as near normal as we can get them, but significantly improved from where they used to be. We're getting a lot more treatment available for patients that I can certainly— that I could provide. And as we get cardiologists and gastroenterologists and other rheumatologists recognising and being able to treat the condition, it really makes a difference to, first, people being recognised as having the condition and getting a diagnosis and, secondly, people getting

appropriate treatment that can be life saving.

**Visual**

**White text on a dark blue screen with a green border around it reads ‘In New Zealand, we have a clinical pathway for Ehlers-Danlos Syndrome and this guides the treatment.’ Next to the text, a white document unfurls. It’s titled ‘Hypermobility and EDS New Zealand Guideline 2019. Includes Generalised Joint Hypermobility (GJH) and Hypermobility Spectrum Disorders (HSD). The green border shrinks and the text disappears, and the document rolls up.**

**Audio**

The way forward for New Zealand for Ehlers-Danlos syndrome is more recognition nationally. The Ministry of Health in combination with the Rare Diseases Organisation

set up a committee, of which I was a member, to write a pathway for Ehlers-Danlos syndrome for both recognition and simple treatment options. This pathway is now present in one district health board (DHB), and we'd really like to see it in all DHBs. We'd like more recognition amongst colleagues of the various issues that are associated with Ehlers-Danlos syndrome.

**Visual**

**White text on a dark blue screen with a green border around it reads ‘Fraser explains the psychological impact of EDS, how we need to recognise the impact of EDS on people and support their mental health.’ The green border shrinks and the text disappears.**

**Audio**

In Ehlers-Danlos syndrome, psychological issues are common. However, they are commonly secondary rather than primary. So if someone is living in chronic pain, it is a major depressant. It can also cause significant anxiety, especially if it affects the person's ability to earn an income. We do have a lot of help from psychologists and psychiatrists in this field, especially when someone first comes to us and they're in a suicidal state. And while we're dealing with many of their medical issues, we need the psychological help to deal with those psychological issues at the same time. The key is that once you deal with the medical issues, the psychological issues lift and lift dramatically. And so people who have been suicidal when they first come to the clinic, as we sort through their issues, they find that life is worth living again, especially if they're not living in chronic pain any more of if they're able to eat again — food that they weren't able to eat before. So the process of building a person back up from a state of injury can take much longer to achieve than someone without Ehlers-Danlos syndrome, for example. And this can be not just because of the soft tissue injuries or the... ligament damage or dislocating joints, but can also be because of other factors in their life that are affected and other organs' systems that are affected.

**Visual**

**White text on a dark blue screen with a green border around it reads ‘Fraser talks about the prolotherapy/sclerotherapy therapy that is used.’ The green border shrinks and the text disappears.**

**Audio**

We do use the strong agents, which are tetradecyl sulfate and polidocanol, which have been used in varicose veins for decades, and they've been used in soft tissue injuries for about 20 years internationally. These agents are much stronger than dextrose, we found, and so we gave up on dextrose at our clinic many, many years ago. However, they help the body lay down soft tissue, build up the tissue and actually stop people dislocating joints.

**Visual**

**White text on a dark blue screen with a green border around it reads ‘This series of videos describe the experience and treatment of one consumer with Ehlers-Danlos Syndrome (EDS). Three members of the multidisciplinary team (MDT) involved in the care and treatment of this person are interviewed. We acknowledge that a MDT can be much larger than this and the video does refer to the wider MDT as an important aspect of diagnosis, treatment and care of people with EDS.’**

**Visual**

**White text on a dark blue screen with a green border around it reads ‘Owing to the diverse and complex presentation of EDS, Rare Disorders NZ (formerly NZORD) has produced guidance (funded by the Ministry of Health) that will help health care professionals and providers to assess, diagnose and plan care for people with EDS. Such guidance will hopefully promote discussion amongst health providers treating EDS, resulting in improved care and outcomes for consumers.’**

**Visual**

**White text on a dark blue screen with a green border around it reads ‘ACC sourced expert opinion about the evidence-base of prolotherapy being provided by Dr Burling. This expert opinion stated, ‘Clearly there is support for this in the literature’.**

[**https://www.hqsc.govt.nz/assets/Consumer-Engagement/Resources/Prolotherapy-review-letter-Dr-Rabago.pdf**](https://www.hqsc.govt.nz/assets/Consumer-Engagement/Resources/Prolotherapy-review-letter-Dr-Rabago.pdf)**. At an ACC review hearing, the reviewer considered this treatment had been excluded from the ACC prolotherapy assessment**

**Visual**

**A white screen with blue and green text reading ‘Health Quality & Safety Commission New Zealand. Kupu Taurangi Hauora o Aotearoa.’ The blue and green company logo comprises of three thin square blocks with white circles of differing sizes within them. Beneath the text and logo, in black text reads ‘New Zealand Government’.**

Accessible transcript by Able.

www.able.co.nz