

# Richard – A boy with cystic fibrosis

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*Cystic fibrosis (CF) is a severely life-shortening genetic disease that can affect the lungs, intestines, pancreas, sweat glands and reproductive organs. In the pancreas, ducts can become blocked and the pancreas can damage itself, therefore, most patients require supplemental pancreatic enzymes and dietician involvement. In the intestine, bulky stools can lead to intestinal blockage. In the lungs, there is mucous retention, chronic infection and inflammation that eventuate in the destruction of lung tissue.*

Richard is a 12-year-old boy who has recently left hospital after an infection related to his cystic fibrosis. Richard has always had a lot of support from the medical profession since he was diagnosed at the age of 8 months. He goes to hospital every 6 weeks for checks and every 12 weeks for routine stays. When in hospital he can spend up to 4.5 hours per day receiving treatment. His height and weight are also routinely measured to make sure that he is growing well compared to others his age.

After leaving hospital, Richard arrives at his GP for ongoing management of his condition and it is apparent that he continues to struggle mentally and physically. Richard has been under a Chronic Disease Management (CDM) plan and unfortunately utilised his five allocated Medicare rebate services for the calendar year. His family falls under the low socioeconomic bracket and they found it very difficult to pay gap payments with the allied health services and have stated they cannot fund further allied health treatment which Richard would greatly benefit from.

The Be Inspired Foundation is contacted to help out with Richard's case. The GP requests that he be able to access psychology, exercise physiology and dietetic services.

*The Be Inspired Foundation with appropriate funding would be able to help Richard by providing him a sponsorship to receive the following:*

## **Psychology services**

- *Interventions*
  - *Cognitive behavioural interventions to improve adherence to nutrition and/or other treatment*
  - *Behavioural interventions aimed to improve psychosocial adjustment*
  - *Cognitive interventions to improve adherence or those associated with decision making in lung transplantation*
  - *Community-based supportive psychotherapy*
- *These services could be provided every one or two weeks, depending on the patient's ability to access treatment.*
- *Treatment would ideally be completed in a group setting to help reduce delivery costs and to increase the patient's social and support networks.*

### ***Exercise physiology services***

- *Interventions*
  - *Aerobic, strength and anthropometric testing to establish a baseline of patient lung and muscle function as well as to monitor progress*
  - *Aerobic based exercise prescription (swimming, cycling and running) to reduce the rate of lung function decline and improve quality of life*
  - *Strength based exercises to benefit bone mineral density and muscle development*
  - *Break down barriers to the incorporation of exercise into a cystic fibrosis therapeutic routine*
- *These services could be provided up to 2 – 3 times per week, depending on the patient's ability to access treatment.*
- *Treatment would ideally be completed in a group setting to help reduce delivery costs and to increase the patient's social and support networks.*

### ***Dietetic services***

- *Interventions*
  - *Monitoring of physical activities and eating habits, and assessment and modification of energy requirements based on growth and weight gain*
  - *Tailoring dietary guidelines for specific patients especially female patients with a potential for delayed puberty because of malnutrition, patients with diabetes mellitus and patients with liver disease*
  - *Titrating salt intake to necessity*
  - *Assisting with nutritional supplementation in the form of either high-energy oral preparations or enteral feeds via nasogastric tube or gastrostomy if indicated*
- *These services could be provided every one or two weeks, depending on the patient.*
- *Treatment would ideally be completed in a group setting to help reduce delivery costs and to increase the patient's social and support networks.*

The Be Inspired Foundation funded Richard's care with a scholarship, as coordinated by his GP, for twelve weeks in total. The psychologist saw him five times in total and assisted him by providing coping strategies to deal with the uncertainty of his disease, cognitive strategies to assist in treatment adherence and conducted a family meeting to discuss the impacts of Richard's disease on his family. The exercise physiologist provided a structured exercise routine as well as seeing him twice a week for a total of 24 mixed aerobic/strength sessions. The dietician was involved in Richard's care four times to conduct a baseline assessment, provide a nutritional plan and manage his ongoing needs.