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One Child's five year flight to Live

By Adrian McGregor.

"Princess Elmira", said the Prince of Light. "While you are sleeping let me show you what a wonderful place my kingdom of heaven is.

"do not be afraid me Princess, I am your friend", and he held out his loving arms to her." Come. I am going to take you for a ride through the heavens to show you my many mansions and houses where I have prepared a place for you to live. Many princesses like yourself already live there..."

Elmira Donaldson, five-years-old, sat in her hospital bed. Listening to the tape recording of her aunt's inspired vision of what heaven would be like for her. Elmira knew she was dying, from cystic fibrosis.

Her parents Jill and Des, had explained that to her the previous morning. But it was a difficult concept for her to grasp. "You don't really die," Jill had told her. "The loving part of you, the real you, the part that thinks and feels, the spirit... You will step out of your body and go to heaven with Jesus."

The next day, Des's sister Delma, arrived at the hospital and unsolicited produced her tape. The Donaldson's listened to it in another room. It broke their hearts, but they realised it was just what Elmira needed. Twice her parents asked her if she wanted to listen to it. "No!" she replied firmly.

They played it anyway, but she ignored it. Why? Elmira Donaldson was a fighter for life. Even in her short five years she knew she suffered to live. Yet here were her parents, who had fought, it seemed, for every breath that Elmira took, asking her to listen to a tape in which it was accepted that the flight was coming to an end.

But the next morning, Sunday, she suddenly asked; "Mummy play Del's tape." She listened, enraptured, and when it was finished, smiled and said. "that's beautiful."

Two days later Elmira died. Not for the Donaldsons. They have photographs of her, tapes of her voice. Her room remains as it was, full of clothes, toys, paintings and the colours of a kindergarten. Outside her window the pink blossoms of Melaleuca eureka shrubs bloom as they did when Elmira lay and admired them.

Her father knows she lived five days, three months and six days. There is nothing maudlin about it. Five months after her death her parents have no reason to try to sweep her memory out of their lives.

The director of the Respiratory Research unit at the Royal Children's Hospital, Brisbane, Dr Paul Frances, remembers Elmira well. She was one of his losses, one of only three in the past 15 months since he took up his post at the hospital. In clinics attached to Australian children's hospitals, 80 percent of cystic fibrosis children now survive at least until 18. Their survival rate subsequently is unknown because new management methods have only recently increased life expectancy for cystic fibrosis patients. A decade ago it was 10 years of age. The most contemporary figures, from Toronto, Canada, have 80 percent of patients surviving until at least 30. ten years hence it may be 40. The improvement in treatment of cystic fibrosis is one of the untold successes of modern medicine, precisely because it is so recent. Elmira Donaldson had severe cystic fibrosis and was not typical of the majority of people with the condition. The National Times acknowledges, but does not concur with, Dr Francis' reservations about the possible negative effects her story could have on the

cystic fibroses community. The opposite side of the ledger is that Doctors thus far cannot detect a cystic fibrosis carrier, do not know the biochemical cause of the condition and cannot effect a cure. Cystic fibrosis is the most common inherited disease in Caucasian children, and the fact remains that 20 percent of those born with it do not reach adulthood.

Elmira Donaldson was one of those. The death of an innocent is inevitably tragic, though the nurse who saw Elmira through her last days said later the scene was as she had always imagined, or hoped, it would be for a dying child. A preparation, no dramatics, an acceptance, no switching off, a belief in happiness hereafter for Elmira, and normal grief thereafter for the parents. But as sad as was Elmira's dying, her life was not. Nurtured in a home rich in sacrifice and succour, Elmira grew a remarkable child to all who met her.

There is nothing superhuman about the Donaldsons, indeed the contrary is what makes their experience worth retelling. "Elmira was a wonderful, brave little girl," said Jill, "She never said she didn't want to die. That's why I don't mind talking about her."

Elmira Angela Jill Donaldson was born on June 17, 1975 at Boothville maternity hospital, Brisbane. Just over 7 lbs, a fair, slight baby, she was a sister to Nigel, who turns eight this April. The name Elmira, Arabic for princess, was the product of Des' scanning an Australian Women's Weekly lists of baby names. But it took several days for the family to accept his exotic choice.

The first five months of Elmira's life were almost as exhausting and frustrating as the last five. In classical medical terms, Elmira failed to thrive. She had bouts of projectile vomiting and was very pale. At eight weeks the local maternal and child health centre sent her for blood tests, which proved normal. The Donaldsons' local doctor could not diagnose the problem. Weeks went by and tension grew within the family. "There's something wrong with that baby's stomach," Jill's mother told her after Elmira threw up a special egg custard the grandparent had made.

Elmira's first visit to the Royal Children's Hospital came when she was three months old. She underwent a series of x-rays and tests and the Donaldsons were told to bring her back in a month unless there were further problems. They were back one week later with their pale, frail baby which had thrown up all five feeds that day. The hospital admitted her, but the very next day there was an outbreak of measles and chicken pox in the children's section and Elmira's ward was shut down. Everything was removed, and the ward fumigated. By an unkind coincidence, Elmira chose her one day in hospital to retain food. She was discharged.

About this time, after three months of constant concern with no answer, Des and Jill began to wonder at their own state of mind. Certainly, when Elmira improved on her one day in hospital, there was a feeling that perhaps part of Elmira's problem was her mother's over-anxiety. But Donaldsons had the fact of their failing baby. They bought scales and discovered she was actually losing weight.

Only Parents who had reasonably ill children can appreciate the bewilderment which now began to overtake the Donaldsons. The act of birth delegates to parents the responsibility for another's health. Failure to sustain that task, apart from their natural love for their infant, attacks the very fabric of their parenthood. One day Jill, in tears, rang her specialist's secretary and told her, "My baby is dying before my very eyes. She is fading away. She is nearly five months old and only 9lb. How far do they let a baby go.'

Back to the hospital for bowel biopsy, liver biopsy, and blood tests, blood tests, blood tests. Jill recalled that day clearly. The hospital staff preferred Jill to wait outside while they obtained blood. "She'll be all right," a doctor told her. "But when she comes out she will be very pale." And she was deathly pale, said Jill. "When I got her home and her nappy off you couldn't count the pock marks down her groin and thighs trying to get her arteries. " But the final test, for the salt content of Elmira's sweat, though painless, produced the most painful result. It is the definitive test for cystic fibrosis, an abnormal salt loss.

Medical folk-law has it that in old Europe the village witch spoke of the child that would not do well if tasted of salt when kissed. Modern clinics see cystic children whose shoes have salt lines on the leather inside from salt loss. He said it was a hereditary disease and that she and Des should have genetic counselling. He would leave it to her doctor to explain. The chief of the hospital cystic fibroses clinic advised the Donaldsons Elmira would live at least until she was 20.

Thus, after 22 weeks, Elmira's condition was discovered by tests that could have been performed at six weeks. Des had never heard of it, or he had, but knew the name and that was all.

Jill contacted the Cystic Fibrosis Association. The very existence of the association says much. Whenever medical science, forever under siege from new and perplexing diseases or problems, fails to provide an answer, the community establishes its own supportive group for those affected. Thus it was with cystic fibrosis, or CF, as it is referred to in association literature.

Des and Jill discovered that had struck the one in 2, 000 chance of having a cystic fibrosis child. Those are the odds.

It is quite a large proportion. Every VFL football team could harbour a carrier. Every school classroom is likely to have one "You might be one," laughed Jill, pointing at me.

Its characteristics are most obviously, as Elmira displayed, the failure of the pancreas to produce the correct digestive juices (enzymes) in the small intestine to break down food. Since Pathologists first described the disease this is where it derived its name – from the cysts and increased fibrous tissue which impairs the pancreas. But far more dangerous is the chronic lung infections which accompany the disease. This is caused by abnormally thick, sticky, mucus in the lungs which harbours staphylococcus and pneumonias bacteria if not cleared. Most people clear their lungs of mucus easily as a constant process of breathing and coughing, CF mucus is too thick to be so cleared. Repeated infections scar the lungs. Eventually it can reduce the oxygen exchange area to the point where respiration fails. Of those who die from cystic fibrosis, most die through lung failure. The key to surviving with cystic fibrosis is clear lungs. Cystic fibrosis is a historically new disorder in medicine. It was first diagnosed in the late 1930s, before which most children died of lung disease or malabsorption of food. Even in the late 1950s well after the arrival of penicillin drugs, the life expectancy was early to middle childhood.

It is only since 1968, with the availability of a wide range of antibiotics and an appreciation of the importance of prompt and prolonged treatment of lung infections, that survival rates have improved dramatically. Because it is not contagious, because it is relatively new and because antibiotics have reduced its ravages, it is not a disorder of which the public is aware.

Elmira's family doctor was aged 60, and he has never before been presented with a case before. De Peter Phelan, director of the Department of Thoracic Medicine, at the Royal Children's Hospital, Melbourne, estimates that the 1, 400 family

practitioners in Victoria have only a 40 per cent chance of seeing one cystic fibrosis patient in their working lives.

“Not surprising most general practitioners will not make a positive diagnosis when they are first confronted with a child who has cystic fibrosis”, he says. But he acknowledges that parents become angry at the failure of doctors to recognise the symptoms.

“I similarly get very angry with my colleagues when it seems that typical cystic fibrosis is missed for many months. The agony parents go through during this period remains with them for the rest of their lives.”

The Donaldsons experienced that agony and in consequence developed an ambivalence towards the medical profession. They grant respect where it is due, equate the merit of a doctor with that of his work.

With Elmira’s diagnosis another puzzle was answered in the Donaldson’s life. They had lost their first baby after three days while living in Nigeria in 1972.

Their doctor subsequently diagnosed the cause of death as meconium ileus, a blocking of the intestine in new born babies. It is now known that 10 per cent of children with cystic fibrosis, developed this symptoms in the first 24 hours after birth. It took a week for the magnitude of Elmira’s inheritance to dawn upon the Donaldsons.

They are an intelligent couple. Both Brisbane born, Des 39, graduated in architecture from Queensland University; Jill, 36, worked as a secretary. They presently run a large squash centre which Des designed and built in Morningside, a south –eastern suburb of Brisbane.

Des is shot, wiry, quietly spoken with a capacity to quickly accept and apply new information. At one stage during Elmira’s treatment a specialist remarked to Des, after Des had skilfully presented a set of symptoms and his conclusions, ‘right, into third year of medicine for you.’ It is said that he wore the final tragedy of Elmira on his face more clearly than Jill.

Jill is attractive, more extrovert. She has a retentive memory and an ordered mind. Staff at the hospital and Elmira’s pre-school, two areas where dedication is accepted as the rule, remain amazed at Jill’s devotion to Elmira.

When I first met they were both in squash kit, a pair of fit Australians. They compliment each other and did so in recounting Elmira’s life to me. This they did in the lounge room of their apartment which Des incorporated in the design of the squash centre.

They needed every bit of compatibility to weather Elmira’s five years. The management of serious cystic fibrosis, as Elmira had is an occupation in itself. The price of Elmira’s health became eternal attention.

First there began three physiotherapy sessions a day, administered by Des and Jill on instructions from the hospital. Twelve positions, in which the principal mode was to lie Elmira on her stomach over a cushion with her body inclined down, and percuss her back with cupped alms to loosen the phlegm and mucus in her lungs. It took half an hour and had to be administered before meals.

Second, Elmira began taking enzyme supplement powder to replace the digestive juices that her pancreas was not producing. In fact it is powdered raw pig’s pancreases, brand-names Viokase. On both parents word it smells and tastes absolutely terrible. To offset this Jill mixed it with strained apple juice, so Elmira ate two spoonfuls of food to one of Viokase and apple. She began vitamin supplement, E, C, K, and more, and of course salt tablets to replace what she lost in her sweat. Since fat is one of the basic food materials most difficult to break down Jill had to use

special oil, known as medium chain triglyceride to make Elmira's butter, to use in biscuits and cakes she baked.

Because chest infections posed such a threat to her, Elmira also began a daily course of antibiotics, on the sound medical premise that it was better to prevent than cure.

She also began a 15 minute session before physiotherapy with a nebuliser. The nebuliser is a mask with an electric compressed-air pump attached which converts liquid into a mist for inhaling. The liquid is Ventolin, brand name for Salbutamol, a bronchodilator, used by asthmatics.

Thus it went nebuliser, physio meal with Viokase, vitamins, antibiotics and salt tablets. That became Elmira's thrice-daily routine for the rest of her life. In the beginning Des had to draw up a check list to remember it all. The treatment would have cost a fortune had it not been provided free as a public ward patient under Queensland's free hospital scheme, for which they were very grateful.

The enzyme replacement program had an astounding effect on Elmira's health. From being 9lbs at 22 weeks, she nearly doubled her weight at 29 weeks. From then she grew at a healthy rate, and she grew stronger her exercises made her a lively little girl. Jill learnt tricks from other mothers, such as the value of tickling Elmira and throwing her in the air to make her laugh, breathe deeply and keep her lungs clear.

There were some changes. As Elmira grew they physiotherapy sessions extended to an hour and Des, with the help of the CF association, bought her a trampoline to bounce on for 15 minutes a before the percussion to help loosen her chest. She transferred to Viokase tablets and consumed 30 to 40 tablets with each meal. "She ate more pills in a day than I have had in my life," said Des.

There was a relief for Jill in a visit to Australia by a Canadian specialist, Professor Henry Levison, of the Hospital for sick children, Toronto, who espoused a normal-fat diet because it helped build up body weight, which was essential when it came time to fight off lung infections. So Elmira waded into milkshakes, chocolates and potato chips, all the good things of a life for three-year-olds.

The next three years of Elmira's life were basically trauma free. Elmira's needs prevented the family waking and making spontaneous decisions for weekends away, but the Donaldsons were delighted with their little girl. She had golden brown hair and was slight from her father's side of the family. Her cheeks her parents referred to as chops they were so large, and she had a smile to match. Her lips were so ruddy you would swear she wore lipstick. Despite Elmira's great health Jill was not deceived. When she first attended the Children's Hospital special cystic fibrosis clinic, she was warned she would see children there who were quite ill.

So she did. She attended four weekly clinics and noticed past surges and declines in children's health depending upon the state of their lungs. The visits served as a warning to be vigilant if she needed such a thing.

At home the Donaldsons fell into a routine which centred around Elmira. The physiotherapy session Des explained it, were simply time consuming and onerous.

Elder brother Nigel, only five, took the brunt of this exacting routine with Elmira. Her needs not only turned the Donaldson's home life upside down, it made inroads into the time and attention his parents had for him.

He was naturally jealous. At first he had bouts of rage where he would tear her books and throw toys at her.

But later he grew out of this and understood at what cost Elmira obtained her attention.

The key to Elmira's development was the extraordinary adult stimulation she received due to the physiotherapy sessions she had three times daily. For an hour either Des or Jill would pound away in the daily dozen positions and converse with or read to her. It became a problem for the pummelling parent to find new material to keep her interested. Aged two she could carry on a conversation and her shelves became full of her children's books. She was quick and perceptive and adored brother Nigel. Later when they played cards together, fish or grab, she would sometimes win game after game until Jill would call a halt to save Nigel's temper. 'One more game, Mummy,' requested Elmira, and Jill would watch her deliberately let Nigel win.

She was a blithe spirit about the house and it infected all. Once Jill broke a glass on the kitchen and Elmira called out cheekily from her bedroom "Of what description?"

She was also inquisitive about her own condition and demanded it be explained to her. They read to her from a Life Science library book called *The Body* and another entitled, *About Me* by Childcraft. From these she learnt about oxygen exchange area in the lungs.

Early in 1978, aged nearly three, Elmira began attending Lady Gowrie child centre, a federal funded, first class pre-school in Fortitude Valley, in the inner city.

The staff were confronted by a pixie faced, pale skinned, fey little girl who was advanced beyond her years both in agility and intellectual ability.

"The first time I saw her she climbed a tree trunk, grabbed a rope hanging off and swung out on it," said Lilac Kendall, the centre medical sister. "I began running, thinking I'll be there to pick her up."

Most children her age could not have done it but Elmira swung calmly back. Child's play to the tyro of the trampoline.

Betty Cassidy, a teacher, remembers Elmira's exceptional conversational powers. But there was more than that. She was an organiser of others and a leader in groups.

Said Betty, "Three-year-olds notice everything. They picked up there was something different about Elmira. Children at that age can be very cruel.

Noticeable about Elmira was her persistent rasping cough, alarming to other mothers until Jill explained it was not contagious. And then there was the gradual clubbing of her fingers, a slight curving and thickening of the blood. If she missed a few weeks of pre-school when she arrived she would be rebuffed by her group. She had ceased to be part of their routine. "But she had this unbounded confidence," said Betty Cassidy. "There was no thumb sucking. She would bounce right back, breeze through their opposition and re-establish herself again."

During her first three years Elmira occasionally had small colds. Immediately the clinic would change her antibiotic, or give her three at a time, anything to wrong foot infections.

In April 1978, just before her third birthday she developed a small chest infection. To be on the safe side she was admitted to hospital for a week for intensive physiotherapy and antibiotic treatment.

This was her first stay in hospital which would become her second home. The children's is beside the main hospital in not unpleasant grounds opposite Victoria Park, a couple of kilometres from the city. Elmira's ward, Turner Ward, was where all cystic fibrosis children stay, was named after the first resident medical officer of the hospital for Sick Children 18189. There was no air conditioning. On a normal steamy,

summer Brisbane day sticking plaster drifted loose from the children's perspiring skins.

The Donaldsons were disappointed at this turn but not concerned. "We would like to have avoided hospital," said Des. "But we didn't see it as a backward step. She was bound to catch some infection eventually."

They prepared Elmira for the experience, explained the reason, kept her at home that night so that there appeared no anxiety on their part and took her to the hospital the next morning. It was a process they were to repeat in the next two years.

Elmira was well and out again in a week, infection cleared. X-rays taken while there showed what staff called "some peri-bronchial thickening." Des understood this to mean scarring of the lung tissue, but he was given no cause for alarm, even when it was repeated in later visits.

In fact it was the continuing diminution of the oxygen exchange capacity of Elmira's lungs, a path that could only lead to one end. Looking back Des can see a pattern of communication with the doctors that he would read differently today. If the doctors specifically said there was no cause for concern, there was none. But if they were confident they could reverse some deterioration in Elmira's health, and if Des did not inquire, no explanation or prognosis was volunteered. It was a part of their positive philosophy program for treating cystic fibrosis patients.

The rest of the year Elmira was well but in early 1979 she went in for another week. This visit they fed her antibiotics intravenously for the first time.

It was effective but a new experience for Elmira. Medical terms can disguise patient discomfort or painful process. To administer intravenous antibiotics a blood sample was taken, the antibiotic introduced and soon after a second sample taken to determine the level of drugs in her blood.

Once, the second sample was taken before the antibiotics had been fed in. A third was necessary. I repeat the error not in denigration - it occurred just once - but to see illness from a three-year-old's point of view and why she cried.

Twice more Elmira required hospital stays in 1979, for four weeks each time. Her height was normal, but she was growing into a slender little girl.

Bearing in mind the dictum that she needed body weight to fight infection, on her last visit that year, in August, the hospital decided to feed Elmira intravenously, a process called hyperalimentation. They made several attempts to insert a cannula into her arm vein, without success. Finally the unit register came to Jill and said, "Look at her weight. It's not good enough. I want permission to go intravenous and I want to give her an anaesthetic to make sure they get in."

Administering anaesthetic to a patient with Elmira's lungs was relatively contraindicated, but it would not be the last time the hospital found itself in a dilemma over how best to help Elmira mend.

In the even Jill signed the anaesthetic consent. The cannula was inserted but within 24 hours Elmira developed phlebitis, inflammation of the veins. It had to be removed because of patient pain. That too was a new experience for Elmira. The next month Dr Paul Francis, now 32 took over had head of the respiratory unit. A young faced, quietly spoken, thorough doctor he trained with Dr Peter Phelan in Melbourne. He is credited with bring alternative techniques and drugs to the Brisbane respiratory unit.

He discovered he had inherited, in Elmira, a child in whose lungs had already detected the more dangerous pseudomonas bacteria. Pseudomonas is a notoriously resistant organism by nature.

There are no oral antibiotics truly effective against it, compared with half a dozen effective against staphylococcus. While it was cultured from her sputum it was of the pathogens which contributed towards the continuing deterioration of her lungs.

Dr Frances was able to introduce Elmira to Cimetidine to aid her gain weight. Cimetidine is a product which inhibits the secretion of the stomach acids which were tending to attach the Viokase enzymes before they could break down her food. It was a new development. The management of cystic fibrosis was actually changing, improving even in Elmira's brief lifetime.

That month, September 1979, soon after Elmira's fourth birthday, the Donaldsons decided they needed a holiday. They chose to go camping at Carnarvon Gorge, a beautiful national park 600 kms north west of Brisbane in the Great dividing range.

Elmira would have to do with out her electric Ventolin mist machine. "But we thought the fresh air and exercise would make up for it," said Des.

As luck would have it Elmira spent the fist three days sick with measles and they ere all on the point of coming home, had it not been raining so much they could have driven out anyway. Yet in the midst if this apparent adversity, the trip became one of the highlight of Elmira's life. They took her on bush walks, Des carrying her on his shoulders when she tired, and kangaroos nuzzled food from her palms.

After a week they moved to Rainbow Beach, just north of Noosa, for a week by the surf. That too was a great success, but it was the bush which caught Elmira's imagination. "Daddy can we go back?" she would often say later. It was like an affirmation of all nature of her, of birth and brightness in the shadow of her own frailty.

In the second term of 1979 Elmira missed five weeks pre-school and by the end of the third term managed to attend only four days. Sister Kendall showed me the A for absent columns beside her name. And when she attended her brightness simply the more contract her debility.

"I used to watch her closely when she came," said Sister Kendall. "She couldn't quite maintain the pace she set herself. I watched her sitting in groups and she would be panting slightly. You wish you could take the next breathe for her." Sister Kendal became lost in thought a moment. "She was never defeated that kid, I really admired it in her."

In February, 1980, a gastric wog put Elmira in hospital briefly, and there was an ominous sign when she was discharged, a slight rash. Jill wondered what it was and it went. In March the Donaldsons clan cleared out for another two week sin Noosa. Elmira looked well and stayed like that to July, gaining weight well and such fitness she could run around a football field.

"We had never seen her so well," remembered Jill. "But she was coughing up excess mucus, too much and the clinic suggested we put her in hospital again to clear it up."

Jill was wary of hospital cross infections, but it was the clinicians view that if Elmira's infection was not cleared up it could be dangerous. She was admitted and within days developed the same rash as February – an allergy to penicillin and its derivatives. It was a blow. Elmira was not allergic to gentamicin, one intravenous drug effective against the pseudomonas bacteria, but she was to ticarcillin, the intravenous penicillin derivative specially effective again pseudomonas.

Although ticarcillin was only part of the total treatment, there is thought to be a synergism – an increased effect – between gentamicin and ticarcillin. The withdrawal of ticarcillin meant the hospital's armoury had been depleted.

Somehow it was symbolised the struggle to come for Elmira. Whenever her parents or the hospital moved to shore up her health, a prop subsided to defeat them. At the very moment they reached to secure her life, it fell a little further beyond their grasp.

NEXT WEEK: Elmira's Flight.

The National Times, March 8 to 14, 1981

Section 2

Elmira the final flight

Elmira Donaldson was born with cystic fibrosis, the most common inherited disease in Caucasian children. One in 20 people carry the recessive gene, but it manifests itself in only one in 2,000. Its most dangerous manifestation is chronic lung infection which, if not controlled with antibiotics, can lead to respiratory failure. Although 80 per cent of cystic fibrosis children now live until at least 18, a proportion die in childhood. Elmira Donaldson was one of those. Adrian McGregor reports.

When Elmira Donaldson, aged 5, entered the Royal Children's hospital, Brisbane, in July, 1980, it was her sixth visit on just over two years.

Despite the discovery that she was allergic to penicillin and its derivatives, the hospital was not alarmed at her condition. She was energetic, attended the hospital pre-school during the day, and was old enough now to be familiar with the staff.

She acquired an 11-year-old friend in the ward and at once out in a request for a double bed. When the child in the next bed became too inquisitive about her private conversation with her beau, she said haughtily: "We'll have to draw the curtains so we can't be watched."

Turner Ward, for cystic fibrosis kids, is unlike any other.

When I solved the puzzle of the childproof gates on the stairs, it appeared the ward was a melee of staff, parents and children, all of whom gazed candidly at this visitor, for that is how reality dealt with them.

The black and white terrazzo floor was scattered with toys, books and pastels, the pale green walls stuck with a clown or animal posters. It was strange conflict of the cheerfulness of children amid the joylessness of illness.

Cubicle E in Turner Ward has a wall panel papered with handwritten instructions for CF treatment – IV antibiotics times, Ventolin times, physio times, special points list, don't-forget lists and daily program, beginning with toilet at 7.30am and ending with lights out at 8.45 pm, containing 26 different instructions.

Because of the demands of the condition parents are welcome to assist where they can in physiotherapy and at mealtimes.

It is undivided attention that cystic children receive which distinguishes them. If you pool together a ward of such children, a lot of them excelling in intellectual development as of them excelling in intellectual development as had Elmira it became a formula for occasions of great delight.

They also knew more than their parents thought. They were the more serious case of cystic fibrosis and they formed a tight-knit group within the ward. If a child awoke short of breath, they knew. If a child was removed from their cubicle they wanted to know why. And if a child died it did not matter whether parents tried to hide it – and some did – they learnt about it from each other.

One evening they were all sitting about waiting to have their baths when the subject of death cropped up. “Oh yes, that’s going to happen to all of us one day,” said one boy. At just that moment their nurse appeared from the bathroom and called “Ok, who’s next?” and was bemused as the group fell about laughing.

Even in this elite group Elmira was noticeable. Her father Des was assisting at mealtime for the group one day when in exasperation he told them; “Come on, you kids, stop procrastinating.” They would not know what that meant, said one nurse.

“Oh yes,” piped Elmira. “It means putting it off until later.” If Elmira heard a word and it was explained, she never got it. She displayed this further, recalling the names of trees when Des took her for walks in a park beside the hospital medical school.

After three weeks Elmira was discharged and the respiratory unit director, Dr Francis, advised the Donaldsons: “These periods of hospitalisation are going to increase.”

Said Jill: “He didn’t elucidate, but he must have seen something.” In fact, Dr Francis was not making any prognosis hospitalisation are going to increase.”

Francis saw that Elmira would require more aggressive therapy in the form of more frequent courses of intravenous antibiotics.

Jill Donaldson had asked for a prognosis on Elmira in February, But somehow never quite received one.

It was in the manner of hospitals. When the ward rounds were made there were several students in tow, ward doctors, a social worker and a sister perhaps. There might be six to eight people around the bed looking at Elmira and little chance to ask personal questions about life and death.

But perhaps too the Donaldsons were reticent about receiving an answer. Sufficiently so not to press for one

Then one night soon after, at home, Jill asked Des: “How long do you think she will live?”

“Oh, 12 or 13,” he replied.

“Darling, she will be lucky to see eight,” she said.

He had been hoping, but Jill had developed a trained eye from her visits to the clinic. Yet even she did not know.

In August 7, 1980, the family went to the exhibition, Brisbane’s show, but 10 days later Elmira, tired and listless, said; “Mummy, I’m not very well.” Jill cried a little and so did Elmira. “I knew she would have to go back again,” said Jill.

Des found her breathing and her pulse elevated, both signs of lung weakness. He was staring to get an inkling of Elmira’s true health. Previously he had read these signs as Elmira’s lung capacity being temporarily reduced by infection. Now he considered whether as a result of recurring infection her lung capacity was permanently deteriorating.

And with that came the apprehension that Elmira’s decline might be inevitable. It was a bleak prospect.

Within a family there is a unique relationship between every member. That of father for daughter is one of intense protection, the unspoken surety that he would

enter battle on her behalf. But Des could not wring any staphylococcus by the neck or dash pseudomonas again a rock. His helplessness as a father would become his agony.

Back at the hospital Dr Francis examined Elmira. He paid particular attention to the jugular venous pressure around the throat. Jill knew from her reading that Francis was looking for signs of heart failure, a sign that Elmira's heart was having trouble pumping blood through her increasingly damaged lungs.

Francis sent Elmira out of the room and told Jill: "Put her in and let's see if we can get a reversal, clear up the infection."

By now the Donaldsons had realised that Jill's estimate of Elmira reaching eight was optimistic. They thought of merely the next year when Elmira was due to begin primary school. They wanted her to have that experience, with all the attendant excitement of a new uniform, new friends, new learning.

And then they began to think about just having her for Christmas. That was only four months off but time was becoming too precious.

Jill told Dr Francis: "I don't want her to die in hospital. It's too cold, too impersonal." This was rhetorical, but she read into Francis' silence the acceptance of Elmira's death.

In fact, Dr Francis was treading very fine line between fully informing the Donaldsons and robbing them of hope. The actual prediction of when Elmira would go was not easy.

Despite her poor appearance, the hospital possessed certain therapeutic manoeuvres. There was a chance she could be given another six months or a year of life. Above all Francis did not want wither the Donaldsons or Elmira to drop their bundle. That would defeat the finest physician.

And there was this too. Death was a denial of the very essence of his profession. Apart from his own affection for Elmira, he was not yet ready to concede the failure if the very best medical management he could provide.

The next day, August 21, Elmira returned to hospital. Turner Ward was full, but the staff shifted another child to make way for Elmira so their past relationship with her would remain as a support. They knew she looked ill, but they too reserved judgement until they saw how she responded to intensive treatment. In fact, Elmira had just one month to live.

It was the last month which, in retrospect altered the Donaldsons's perception of themselves and their future. Back in January Jill's IUD failed. She became pregnant.

Elmira was delighted at the prospect of no longer being the baby of the family. Her parents were less so. They were becoming increasingly aware of their responsibilities as CF carriers. The chances of two carriers of the cystic fibrosis recessive gene having a cystic fibros child are one in four. The fact the Donaldsons already had one such child did not alter the odds. With each new pregnancy it was one in four. Des and Jill decided to accept the risk. Jill lost the pregnancy at 20 weeks. They would not make the same decision again now, they told me it was the last month that did it. "Before that I could have contemplated losing Elmira and still risk having another one," said Jill. "But after what we went through in her actual dying... I would never subject another child to the risk of that."

Des agreed. After the first death there would be no other.

On August 21, the Donaldsons began a schedule which was to dominate their every hour. Elmira spent nights in hospital receiving intravenous antibiotics and wearing an oxygen mask as part of intensive therapy.

At 6am Jill would drive from Morningside to hospital, assist with Elmira's treatment and at 10am bring her home. At 2pm they would whisk her back for half an hour of intravenous antibiotics and be home by 3pm. Then, about 7pm it was back to hospital for the night. All this time Elmira had a cannula strapped permanently into her left arm, protected from bumps by swathes of bandages. It hardly seemed her thin arm could carry it. She did better than that. The cannula had a tap attached through which antibiotics could be injected. It saved having to be jabbed half a dozen times a day. Elmira would often press the syringe herself which the ward sister, who had set the equipment up, supervised.

The family had two cars, a Morris 1100 Jill's parents had bought her at Elmira's birth and Des' older Holden. The trips back and forth took to the hospital took the best part of two hours a day. Occasionally, at Elmira's request, Jill would doze down on a banana lounge or lounge chair and stay the night.

Hospital staff who did not know of the Donaldsons' squash centre wondered how Des could manage to spend so much time by his daughter's side. The answer was Jill's parents and Des' mother, who were running the squash courts as Elmira consumed her parents energy and time. It was simply further help for which the Donaldsons were grateful.

All of this change in routine was hardly lost on Elmira. She saw Des taking photographs of her and was sensitive to the subtle changes in people's attitudes towards her. Normally Jill insisted that Elmira's grandparents keep large gifts for special occasions, Christmas or her birthday. Jill's father wanted to give Elmira a Smurf jewellery kit and Jill told him: "Dad, let her have whatever she wants."

Although the Donaldsons girded themselves, they still had genuine hope and spoke about another bushwalking holiday. But Sunday, August 31, at Morningside was an unforgettable day for the Donaldson extended family. Elmira's grandparents were over to watch slides that evening. Afterwards Des gave Elmira a bath and unexpectedly she threw the bath washer at him. "What's that for?" demanded Des in injured tones. "To remember me by?" said Elmira laughing, gazing at him unfathomably. It was effective. "I did a double take," recalled Des. "That was the first sign I knew she knew..."

Des taped some of the proceedings that evening. Looking at her photograph and listening to her a high-pitched childish voice, aggressive, strident, full of ideas and comments, vain that she was being recorded.

"I'll bet I can make you say black," she challenged her grandfather. He was game.

"Name the colours of the Australian flag?" she shrilled. Um "red, white, and blue". "I told you I could make you say blue," she said.

"But you said you would make me say black...oh!" said her grandfather. Shrieks from Elmira. But in between there was this rasping, incessant cough. The her grandfather innocently inquired: "What would you like me to bring you in hospital?" He was in the habit of doing that. Elmira thought for a moment, resting her head against the lounge room wall. Grandfather waited and then persisted, "Oh you don't need anything". But then it came to Elmira, and she replied softly: "A new life."

It was the moment for Des and Jill to start talking to Elmira about dying. They sensed she wanted to. But it was a family occasion. They did not want to morbid. The hospital doctors had not uttered any sentence. They decided to err on the side of positive. In a way Elmira was disadvantaged by that positive policy. Because the

hospital would not confront the Donaldsons with the probability of death they could not bring themselves to face Elmira with it.

Still Elmira's spirits did not flag. The previous week, a second nurse, Margaret Mitchell, 20, had been rostered to Turner Ward. She was like Jill, a born-again Christian, a person who had given control of her life to Jesus Christ. If I had asked with her or Jill to explain such a coincidence they would have replied it was no coincidence, it was God's will.

She had read principals of Elisabeth Kubler-Ross author of *On Death and Dying*, as part of her training. She had strong views on the quality of life as well as the quantity and was please to discover that some of this was practised in the hospital. Jill too had read Kubler-Ross and she struck up an immediate rapport with Mitchell. The friendship proved a support to her and Des in the few weeks to come. But initially Mitchell had to combat the urchin Elmira who immediately pulled the I-can-make-you-say-black trick on her. Mitchell retaliated by fetching her younger sister's 101 elephant jokes book to fight fire with fire. Sample: How do you get down from an Elephant? You don't. You get down from a duck!

As healthy as was Elmira's wit, it was evident she was losing weight, losing vital strength. September 7 brought a traumatic day for her, her parents and the hospital staff as they attempted to introduce a nasal gastric tube to feed her. Elmira was violently opposed to it. Jill stayed that night and the next morning Elmira vomited the tube out. They tried again but could not even get it her throat. After another week, in which Elmira only picked at her food, the hospital proposed intravenous feeding.

Dr Francis knew from Elmira's records that it has been unsuccessful before, but he had a few options. Without it Elmira was becoming progressively weaker, It was an unhappy decision for him and the Donaldsons.

That meant Elmira's sojourns at home ended. When she returned to the hospital that Monday evening she stayed. And never left again. Now, even on someone like Elmira, the strain began to tell.

The next morning she showed how near the surface was her concern at the unknown quantity of death. At the end of one physiotherapy session she was so short of breath she began to gasp. She panicked and cried out: "I'm dying! I'm dying!"

Jill calmed her. Later she asked Elmira how she knew she was dying. "Because I can't get enough oxygen in my lungs," said Elmira, quoting from the medical books she had been read.

Two days later, Elmira's bed was moved to be placed in front of the Turner Wards main desk to be under consent supervision. She began to wear her oxygen mask full time, instead of just at night. From experience this signalled to Jill that Elmira had only a week to live. It was September 18.

Next day, the fourth day of intravenous feeding, unit director Dr Francis asked the Donaldsons for permission to stop it. Fluids introduced directly into the blood stream increased the volume of fluid Elmira's already overstrained and failing heart had to pump, It was causing distress, said Francis.

It had been a risk in which the calculations were that if Elmira did not benefit it should not produce any irreversible condition. The halt, in fact, signalled the hospital's acceptance that they were losing Elmira. It was no small admission. Turner Ward had known Elmira all her life it seemed, a life which they were instrumental in extending.

The moment when the staff came to the belief was not perceptible in their manner, because Elmira was not the only child in their ward, all of whom depended as

much on the cheerful mien of staff as on any drugs, A smile if often the heaviest burden of nursing. But where once, if they thought they could reverse Elmira's decline, they would have pushed on, now they began to tailor her treatment to comfort her as well.

Yet another balance had to be struck. Elmira could not be saved but if they removed the cannula feeding her antibiotics intravenously, Elmira might see it as the withdrawal of her life support systems. The hospital was not abandoning her, nor would they have her fear so. Her medication was simplified, but they kept that cannula strapped to her arm. If the breathless incident a few days earlier had frightened Elmira, it had no less her parents. On Friday night, September 19, they resolved that there was no further point in pretending. They must talk to Elmira about dying.

Jill had twice invited Pastor Clark Taylor, of the Christian Outreach Centre, to their home for healing prayers. Pastor Taylor had prayed that God would make Elmira whole again and that he would take away her pain. "They were a devoted family," he told me. "Loving parents. What more can I say?"

Elmira has some idea of heaven. In hospital she had played with paper cartoons on which transfers of people could be placed to fill in the story. Elmira's cartoon series was Little Red Riding Hood. Elmira had an outfit just like Little Red's. She placed the transfers according to the story until the very last picture. On this she placed Little Red Riding Hood up in the sky, amid the Stars.

Why? asked Jill.

"Because," said Elmira.

On Saturday, September 20, Jill and Des broached the subject with Elmira, Jill carried the burden of explanation of Dying, of spirit, of Jesus and of Heaven.

"When we told her she was marvellous," said Jill. "It was like a load lifted from her shoulders. She had been waiting for us, I suppose. She didn't say she didn't want to die, you know. Not once."

Elmira did ask, "Mummy, I want Annie, get Annie," Annie was a little cystic fibrosis friend in the ward, six months older than Elmira. Jill brought her to Elmira's bedside. Elmira took off her oxygen mask, put her arms around Annie, pulled her face down and gave her a long beautiful kiss. Then she put her mask back on. No words were spoken, but her embrace was more eloquent than any farewell.

From the previous day either Jill or Des stayed at the Hospital 24-hours a day. At first while she wore her mask they could talk to her, read books, or watch colour television. But by that Saturday she had begun to lose interest in play. The days were still full. Jill brushing and combing Elmira's hair, physiotherapists returning every hour and a half for short sessions.

That Saturday afternoon, Nigel and Jill's mother and father came to the hospital to say goodbye. Not in so many words, but the grandparents had not seen her for a week. The deterioration was a shock to them.

It is a continual process of cystic fibrosis that air, trapped in the lungs because of the damage to the alveoli, lung cells, causes the chest cavity to enlarge. Jill's parents saw a little girl with thin arms, round shoulders and a barrel chest struggling for breath even through her oxygen mask. They knew they would not see her again. It was heartbreaking. They cried, Jill cried and Elmira cried too. But then Jill pulled herself together, turned her tears into a laugh and said: "Come on, Mummy can laugh." Elmira brightened and did not cry again.

On Saturday afternoon, Nurse Margaret Mitchell came back on duty after two days off to discover a startling decline in her young charge's condition. On Wednesday she had dared Elmira that she could not eat five spoonfuls of ice-cream. Elmira had eaten six, turned to her mother and mock-complained "She made me eat that." Said Jill: "She should have made it 10."

Mitchell had left at 3.45 Wednesday afternoon, and when she returned at 2.45pm Saturday. Elmira could manage only a wan smile for her. No jokes, no clash of words, just a weary little girl. It was a dramatic downturn.

After Jills' parents left, Des's sister, Delma, arrived with a tape of what Heaven might be like for a five-year-old. She had been disturbed the previous day by Elmira expression her fear of dying, of the unknown. "I think you should listen to it first," she told Des and Jill. They did, and it reduced them to tears. In its way it is a masterly mixture of child psychology, adventure tale and Christian hope. It draws in part from Revelation, but it is mostly Delma's inspiration, related to Delma's broad Queensland accent:

"The Prince of Light took Elmira through field of flowers that smelt like a world of roses. Mmmm. He let her play in the lush green roly grass and slide down rainbows and play with all the other kids. She ate and drank the best food she had ever tasted. Elmira dressed up in the finest soft gowns of pink and yellow and put sparkling jewels on her fingers and a beautiful diamond tiara on her head and crystal slippers on her feet just like Cinderella's. How beautiful she looks."

The tape ended with the lilting piccolo with signals Morning from Peer Gynt by Eduard Greig. Elmira knew the music.

When it finished, Elmira and her parents talked again about dying. Said Des: "I think that inside she was a bit frightened, I really do. But now she had something to hang on to." Jill concurred. "Elmira had a greater acceptance of what was going on after that," she said. If anything, the Donaldsons blame themselves for not talking to Elmira sooner. "When we did, it was too close to the end," said Des. "Back at the slide show evening, when she asked for a new life-that's when we should have done it. Before she became distressed."

After that they did not talk about it again. "We just left it," said Des. "We didn't know what she could cope with. We weren't experienced. We didn't know what to do really, other than try not to distress her."

Elmira accepted her parents' explanation of death, founded upon Jill's profound Christian belief. But there were nuances to the rationale which an intelligent child like Elmira could not miss. There were questions which were divorced from the straightforward fear that death would separate her from her mother and father. Elmira's whole life had been devoted to maintaining her health. That in itself instilled in her the intrinsic value of life. The positive side of her condition, which gave her suffering a form of purity, had been the pursuit of her very existence.

However much her parents gave her the promise of heaven, Elmira experience, which had so emphasised life, almost precluded any alternative. Later that Saturday Elmira let forth a long sigh and exclaimed; "Oh, dear, Oh dear, Oh dear."

"What's the matter?" asked Des.

"This is the end of my life," said Elmira quietly.

Des didn't know what to say in return, it was like an adult had addressed him. "Well it's not yet," he said and began reading a book to her.

Next day Elmira suddenly said; “Mummy, I feel strange. I feel like my head is over here and my arms are over there.’ Jill’s mind flew to religion, that Elmira was about to die, that her spirit was leaving her body.

‘Are you going, are you going now?’ she asked Elmira anxiously. That was the phrase they used, going to heaven.

“No,” Elmira said. And it passed. They had no answer but it showed then she understood some concept of going.

But Sunday began Elmira’s Trail. Her physical distress began to overtake all other considerations. Her life revolved around the oxygen mask. She was most comfortable sitting up, lying forward over a pillow. More and more she wanted to get into that position. Lying back was not comfortable.

Some time during Sunday Des took his daughters pulse. It was 150 constantly, and she was taking 50 breaths a minute, nearly one pant a second. Once she cried out in anguish: “Oh, stop, stop.”

‘What darling, stop what?’ asked Jill anxiously.

Her heart. She wanted her heart to stop, said Jill. It was draining her. “Her body had taken control and was making her work hard,” she said.

There were little the hospital staff could do and they were not immune to the emotions being engendered in C Cubicle. It had been almost cleared of other patients to give the Donaldson privacy.

Before she finished her Monday shift, Nurse Mitchell asked to be assigned to Elmira the next morning, Tuesday. But she was refused because she would be too upset. As it was, she had time to tiptoe in and she was upset. But others’ tears can be a comfort and they were, and she was, to the Donaldsons.

Some days previously, Elmira had bouts of withdrawal from her parents. “Don’t touch me,” she would scream, as through they were anathema to her flesh. But on Sunday she needed all the comfort they could give her. She had pain in her stomach which Des eased by holding her.

The process of breathing has become a terrible struggle. She drew massive breaths, her chest heaving out and shuddering back, and her stomach muscles working in unison. Her whole body involved in the effort of breathing and after bad spells her head and body would be lathered in perspiration.

She had not eaten for days and the effort was slowly consuming her. Her lungs now produced mucous so viscous than when she coughed it came up like thick brown treacle.

Thus passed Monday, September 22. Elmira endured it without complaint. Although at one stage she did day to Jill: “Mummy, I want to go home.” She knew, and Jill knew, she could not, but it was what they both yearned for. To go home.

That evening Elmira had a slight remission. Her breathing subsided and she rested more easily. “She picked up,” said Jill. “It was strange, wasn’t it, Des? Maybe I was hoping for a miracle, but I thought that perhaps she had gone so close and maybe...”

Both parents thought the same thing. In a moment of joint hope, Des said: “Wouldn’t it be lovely if she got better from now on?”

But the distress returned, Elmira throwing her body around, on and on, until Jill it seemed it could not last. Des took the shift until midnight and was so tired he lay down on the bed next to Elmira’s. Jill took over but, exhausted put her head down on the bed and dozed until 4am...

“Elmira!” – they were awoken by a nurse’s shout. Elmira was standing beside her bed, calm, her breathing natural and even. The contrast with her previous distress was incredible. Des gathers her in his arms, her nightgown flowing down, as Lear carried Cordelia, and put her back in bed. She took a long drink of water and bed noticed that the tips of her fingers had turned pale blue. Lack of oxygen to her extremities. And he understood her body had given up. The mask could supply oxygen to Elmira, but her lungs were so damaged, so depleted of space, she could not breathe well enough to adequately oxygenate her blood. With her gas exchange area diminished gas levels in her blood were altering. Oxygen down, hypoxia and carbon dioxide up, depressing her central nervous system. Elmira was subsiding into a state of carbon dioxide narcosis. It was probably the best thing that could happen. Now it was a matter of how long she could survive with that depleted oxygen flow.

Jill. Exhausted, fell asleep again, while Des kept watch. At 6am he saw the sky grow blue outside. He leant forward over his peaceful daughter and said softly; “Look chicken, it’s a new day.” Elmira actually looked at the sky outside. That was the last living recognition she gave him. She fell deeper into a coma. Des woke Jill and said: ‘I think it must be getting very close.’ Both parents sat in vigil by Elmira’s bed. Around them Turner Ward rose to meet the new day, but no-one intruded upon them. A nurse put a stethoscope to Elmira heart. It was very slow. Her breathing grew quieter and her pulse more faint, until 7.40am she became more silent than life. Elmira had sought the relief she had sort. Her heart had stopped.

“Then magically, as if in a dream, off we went into the blue sky. Elmira looked down. Gosh, she said. I can see Mummy hanging out the clothes and Daddy mowing the grass. Yes, said the Prince of Light. From my kingdom you can always watch your family and see how they are from day to day. Where I live there is no sickness, no pain, and no sadness. Only happiness.

“ Looking up. Elmira saw angels open the gates of Heaven. In side was all yellow and softly glowing light of a thousand candles.

“Sitting on a golden throne set a sea of sparkling jewels was the father of all mankind. In his hand he had seven small sparkling stars. He gave them to her and smiling said; “Welcome, Elmira.”

From the Readers.

Elmira’s Story.

Sir, I want to express my appreciation to Adrian McGregor for his two articles, titled “Elmira”, (NT, March 1 and March 8). It was a particularly sensitive presentation, and in among the human story, he gave the facts about cystic fibrosis is as accurate a form as I have even seen in the printed medium.

As you have observed, cystic fibrosis is the most common lethal gene in the Australian community, yet financing of research into the problem is left almost entirely to the parents of the sufferers of themselves.

To the best of my knowledge, not a cent of money from the National Health and Medical Research Council has found its way into the several Australian laboratories performing basic clinical research into disease and its relief.

By contract, the National Institute of Health in the US pours millions into CF research and treatment. Perhaps we just don't have a strong enough lobby.

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