



Update on Nutrition in CF: Zoom on Small

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The title puts in words how present nutritional care in CF surpasses the mere treatment of malnutrition and increasingly zooms in on inconspicuous details like micronutrients or the very small: newborns and young infants diagnosed early thanks to neonatal screening. This article will cover both these items but first pay attention to prevention of malnutrition and to nourishment aiming at a normal physical and mental development.

The Past: Treating Stools

We have come a long way in the short history of cystic fibrosis. CF was originally described in 1938 as an exclusively digestive disease causing early death in infants due to extreme malnutrition before any sign of respiratory illness could develop. Autopsies showed the pancreas to be the culprit: it was full of small holes (cysts), surrounded by fibrous scar tissue. The entity was called “cystic fibrosis of the pancreas”. These abnormalities perfectly explain the clinical picture: the early progressive destruction of the organ causes elimination of its function which is central in the uptake of nutrients from the bowel as it produces the essential enzymes to digest starch, protein and especially fat. Consequences can easily be perceived by the senses. The smell of decomposing protein is characteristic and the amount of fat in the stools increases their volume, makes them look pale, yellowish, float on the toilet water and difficult to flush. This description is rather repulsive but gives important information because it can be performed by oneself and for free. The most important consequence of the loss of nutrients of course leads to failure to thrive.

From the observation that the absence of pancreatic enzymes was the cause of the disease, it was suggested to replace them by an animal surrogate: a powder made from dried, grinded, purified piglet pancreases to be taken with the food. It worked although minimally, because large quantities had to be given in order to obtain small changes in stool appearance. For the ambitious, fussy doctors it was disappointing. They came to the unfortunate hypothesis that it must be possible to diminish the quantity of fat in the stools by allowing less of it in the diet. They proclaimed fat prohibition: fat was banned from CF cook books but as a result, so was taste. Fat free dishes were unpalatable and reduced the patients' already small appetite. Being obedient, patients followed the prescriptions even though they hated them. Unfortunately, it worked: never had CF stools looked so well formed, so healthy as thanks to these commands! However, as Murphy says, "mother nature is a bitch" which does not grant doctors the pleasure of a victory. As the volume of the stools diminished, so did the patients weight. While stools looked better, patients got worse. They developed a hunger profile, recognizable from miles away. Their belly was massively swollen while the rest of their body was very skinny. They had no buttocks, no subcutaneous fat, hardly any muscles, looked pale and sad and had little energy



reserve. No wonder they succumbed at the first serious infection. This was -erroneously- regarded as the typical CF appearance far into the eighties and most of these children remained so for the rest of their short life. In retrospect, it is incomprehensible that the medical world did not see that they inflicted an unacceptably high toll for the shape of stools.

The Discovery of Energy Needs

Fortunately not all CF caregivers accepted this fat prohibition indiscriminately. Some, like good accountants, first compared expenses and revenues by the simple observation of the obvious. People with CF have higher energy expenses due to the disease itself, to the increased respiratory labour, the constant fight against infection and the imposed physiotherapy and sports. They also lose more in their stools as a result of maldigestion and malabsorption, in their sputa which also contain more protein and in their urine when they have diabetes. The addition of all these losses raises energy needs by 20 to 50%. When this excess is not covered, weight gain nor growth can occur and defence mechanisms will be jeopardised making patients prone to respiratory infections. This is the pathway created by fat prohibition since fat is the main energy provider in nutrition (fat has nine calories/gram, which is more than twice as much as carbohydrate or protein). Conversely, the sequence can be reversed when more energy is given and the nutritional state improves, helping to prevent pulmonary deterioration.

This basic analysis made the doctors who valued persons more than stool, critical. They concluded that energy could never be kept in balance without fat in the food; they rejected fat prohibition and, on the contrary, recommended a higher consumption.

It was only years later and by coincidence that the results of this approach became clear. The CF centres of Toronto (Canada) and Boston (USA) closely collaborated by exchanging caregivers and following identical guidelines for respiratory treatment and use of antibiotics. Yet when the data were compared Canadians were doing much better than Bostonians: they were heavier, taller, had better lung function and were surviving longer. Scrutiny of all treatment details only showed a difference in diets: it was fat restricted in Boston and not in Toronto. In Toronto they even recommended more fat. Could this make the difference? Although disbelieving, fat prohibition was progressively abolished. This was a hinge moment in the history of CF, although the basis was humiliatingly simple: people should be in the centre of medical attention, not stools. The resulting lesson is of key importance in the care for people with CF: energy is the magic cornerstone, which should be provided by food, lots of food!

At first sight, this seems easy in a community with a growing number of obese who eat too much. It is however torture when you constantly have to eat more than your appetite, especially when this is suppressed by cough and illness. Increasing the volume of food eaten by 20 to 50% is impossible for most patients. It is more effective to try to increase the calorie content by adding sugars and fat, thus also turning the fat free rubbish into tasty delicacies. This type of food preparation comes close to that for the general population, which we condemn as unhealthy because it causes obesity. For people with CF this is the right food although we are hesitant to recommend the generous use of double cream, fat rich sauces and sweet desserts. Our shame disappears however when we see that patients appreciate it much more than the former fat free concoctions. People with CF are stimulated to become gourmets, dieticians are retrained from nutrient calculators to gastronomic advisors and parents are taught to become "cordon bleu" master chefs. It's not always good for the parents' weight but it's excellent for the patient's physical and mental health. When restrictive diets disappear the social function of eating returns. They can go to a restaurant and, thanks to a better physical appearance, enjoy the company of friends and even consider creating a family.



Behavioral Obstacles

It is unfortunately not all roses. Some patients keep us with our feet on the ground. Nutritional advice is often challenged by the supremacy of a slim figure imposed by lifestyle television programmes and glossy fashion magazines. They make great impression especially on young girls for whom encouragement to eat somewhat more in order to gain some weight is like incitement to sin. These are extremely difficult clients because they are encouraged by powerful advertisement and appreciation for a skinny figure by envious friends. Here we are glad we can call in allies, psychologists who can not only tackle the eating problem but also those of puberty and adolescence in general.

The help of psychologists is also invaluable in behavioural feeding problems at any age but especially in pre-school children. These are not specific to cystic fibrosis but probably more frequent and more frustrating because parents are cornered between an unwilling child and an insisting caregiver who threatens with the deadly dangers of inadequate feeding. The parents' reaction is often inappropriate which can lead to dramatic situations at the table and in the family. The resolution mostly takes a lot of time but the united endeavour of a patient's psychologist and dietician can work miracles.

In recent years more attention has been given to “the side show” of eating. It is clear that the environment and company of a meal can be more important than what is on the plate. This is certainly true for the special circumstance of CF where people have constantly to eat more than their appetite. We therefore have to be very broadminded and allow, even stimulate, food which is discouraged for others as unhealthy. Fast food like pizza or chips, dressings, chocolate, peanut butter and candy bars can at times be helpful resources. After all... the tricks and attempts to enrich regular food can lead the horse to the water, but can't make it drink. Neither can we make it absorb!

And yet digestion and absorption is severely put to the test by the higher ingestion of fat. Even more attention must be given to taking enough pancreatic enzymes with all food. Fortunately pancreatic enzyme preparations have become more potent, thanks to a better presentation and a higher concentration. Yet this does not always resolve all digestive problems. In a vain attempt to reach this, patients have excessively increased dosage of the highly concentrated preparations and thus created a new disease, “fibrosing colopathy”, in which the walls of the right large bowel thicken and eventually even obstruct passage. Thanks to a better control of enzyme dosage the disease has practically disappeared. Meanwhile the value has been discovered of medication which decreases the production of gastric acid and thus prevents rapid destruction of pancreatic enzymes in the stomach. It reinforces the enzymatic power so successfully that it became part of the standard treatment in many centres.

Special Aids

CF is often nastier than the above suggests. At times infections occur during which appetite is very low and yet more energy is needed to fight them... For these special circumstances industry puts concentrated sip-feeds on the market in small brick packs containing easy to take high calorie drinks in many colours and flavours which are very good as in between snacks or supplements after meals. They are very successful in repairing energy balance over a short period but should not become easy sweet substitutes for regular food.

Once the dietary intake becomes problematic and this can be anticipated to be of longer duration a more drastic solution must be proposed: enriched regular meals continue to be given supplemented by hyper-caloric sip-feeds. In addition, a high quantity of calories can be supplied “effortlessly”. This is most conveniently performed through a gastrostomy, a direct opening between stomach and skin through which a discrete silicon device is placed using endoscopy. During the day, when the gastrostomy is closed, all normal activities like going out, swimming, sports, normal eating are possible. At night the gastrostomy is opened and connected to a feeding bag. A large quantity of calories and nutrients is provided in the form of a special formula containing the latest advances in nutritional science, e.g. predigested protein and fat for which no pancreatic enzymes are needed.

Maxi Important Micronutrients

Anti-oxidants are an important result of the latest nutritional research, paramount for CF. In this disease inflammation is at the origin of pulmonary damage. It is initiated by infection and oxidation is its mode of action. Normally it is counteracted by anti-oxidants but in CF these are not sufficiently available since one of the most potent, vitamin E is fat soluble and is not well absorbed due to the malabsorption.

This is unfortunately not true for the most potent water soluble one, vitamin C, which is naturally abundant in fruits and vegetables.

These foodstuffs are however not the most popular for children and medical caregivers insufficiently include them in their nutritional recommendations which have mainly eye for fat and calories. In a recent study our dieticians found out that 20% of our CF patients took less than the normal recommendations while they probably need more. Subsequently more attention is paid to fruits, vegetables and oils rich in anti-oxidants and to the daily systematic consumption of at least 200ml of juice.



Since the earliest years, fat soluble vitamins (*ADE & K*) were supposed to be problematic in CF because of the fat malabsorption. But, remarkably, even though blood levels have been reported to be low, symptoms of deficiency have very rarely been described. To stay on the safe side however a supplement is systematically prescribed, preferably in a single preparation. Vitamin E is given in a higher dosage, for the reasons we described above and because there is no danger in giving more than the daily needs. This is also true for vitamin K, shown to be important in more than the known blood clotting but also central in the maintenance of bone integrity. The cornerstone in this respect is vitamin D for which we recently demonstrated that the sun is much more valuable than oral supplements. Since it is not available during the long dark winters, prudent sun exposure should be encouraged during the sunny months.

Calcium is the substrate for vitamin D; it is the third nutritional brick in the bone wall. It can copiously but almost exclusively be found in milk and dairy products, excellent food for CF since they contain a myriad of beneficial factors, easy to take, at a cheap price and with a multitude of applications in the kitchen. Yet our dieticians found out that they are insufficiently present in the menu of 40% of the people with CF. Patients should know that maintenance of good bone health is of utmost importance since very painful fractures of ribs and spine can occur without trauma. They should be encouraged to drink 800 ml of liquid dairy product a day, which provides the needed 1 gram of calcium. Hard cheese is even richer and can in grated form be added to many dishes.



Essential fatty acids -EFAs- are increasingly shown to be beneficial in CF. At this moment omega 3 receives most attention e.g. in fish oils. The EFA should abundantly be present in a varied CF menu through fatty fish like salmon, halibut, herring, etc. or oil of cole-seed (canola).

Finally, the food of the very small, the newborn and young infant should receive attention. In many regions and countries diagnosis is made at an early age thanks to systematic newborn screening. This is as efficient as the subsequent follow-up and thus only the very best nutrition is good enough. For every infant this is the mother's own milk. Pancreatic enzymes should be given at each feeding session and the infant must be followed very closely in order to detect problems as soon as they appear. When breast feeding is impossible, industry now puts a special formula at our disposal, containing more calories, salt, vitamins and proteins and fat in a pre-digested form, needing less enzyme substitution.

It is worth while...

The provision of nutrition to somebody with CF is not easy. It is demanding and often frustrating to withstand the publicity tsunami of "slim and light is healthy" and to continue the search for the miraculous nutrient which will put more weight on the balance and yet goes down just right. This daily endeavour is however not in vein since it is an established fact that survival mainly relates to the nutritional status. As the latter declines so does survival. This is increasing every year without negative effects on the quality of live and steadily crawls to the final goal for people with CF: the prospect to develop diseases of old age.