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Gastrostomy Tube Feeding of Cystic Fibrosis Patients

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Objective: To identify the implication of gastrostomy tube feeding of Cystic fibrosis (CF) population in Saudi Arabia and its effect on mortality.

Method: A retrospective chart review of all confirmed CF patients for the period Nov.1994- Oct. 1998 for demographic, clinical and nutritional data.

Results: Of 96 CF patients, 81 (84%) CF patients are a live, 15 (16%) died. Fifty (52%) were males and 46 (48%) were females. Age at diagnosis 2.9 ± 3.5 years. The mean follow up period was 3.24 ± 2.8 years. Sixty five percent of patients were in the mild to moderate malnutrition stage (<90th percentile) weight for height percentile(Wt/Ht). Sixty three percent were in the mild to moderate stunted growth (<90th percentile) height for age percentile. Gastrostomy (GT) tube feeding was established in 8 patients, 5 were females and 3 were males. Age when GT inserted was 5.5 ± 2.8 years, a range of 2.6-11.5 years. 2 were still alive at the time of the study and 6 died after GT intervention. Duration of GT feeding was 13 ± 11 months, a range of 1.4-38 months. The weights and heights of all patients with GT improved minimally in the first 6 months, but remained the same thereafter. Comparison of all CF patients who are a live at the time of the study and patients with GT feeding have shown that factors contributed to early mortality of GT patients were: Mucoid pseudomonas colonization at diagnosis (p 0.03) and development of early resistance to Genta (p 0.04).

Conclusion: Early nutritional rehabilitation is necessary to improve the survival of CF patients. GT feeding may not improve survival if placed in patients with progressive lung disease and moderate to severe malnutrition stage.

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The incidence of cystic fibrosis (CF) in Saudi Arabia was reported to be 1 in 4243 children[1]. Epidemiological and genetic data have been described in details in many Gulf countries[2-5]. The mean survival in many Arab countries has bee low, around 10-15 years of age[2-5]. This fact has been related to many factors such as, delayed diagnosis, delayed treatment, Genetic mutations, and poor compliance to treatment. No detailed nutritional data or discussion of any type of nutritional intervention have been discussed before in the Arabian countries[2-5].

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The importance of nutritional status in long-term survival and well being of patients with CF is well documented[6-8]. There is a clear association between malnutrition and deteriorating lung function[8]. Chronic pulmonary infections (particularly with Pseudomonas aeruginosa (Pseud) are associated not only with anorexia, but also increased metabolic rate and energy requirements[6.9.11]. Consensus report and recommendations were generated by the CF foundation (CFF) in USA to provide nutritional guidelines for the care of CF patients[6]. Weight (**Wt**), Height (Ht) and growth velocity are important parameters in the evaluation of CF growth[7,12-14]. CFF recommended that active dietary intervention should be initiated when Wt declines below the established Wt curve or becomes under Wt (85-89%) of ideal Wt/Ht. Nutritional intervention in the form of parenteral nutrition through naso-gastric tube feeding (NGT), or gastrostomy tube feeding (GT) have been shown to be very effective tool to improve the survival and quality of life of such patients[9-15]. Corey et.al.1988 (12) compared 2 CF centers in Toronto, Canada and Boston, USA, with similar CF population size and age distribution, but different nutritional management in that: Toronto center advocated high fat, high calorie diet with up to 20-30 pancreatic capsules/ meal, whereas Boston center advocated a low fat, high calorie diet with less pancreatic enzymes/ meal. He found that, Patients in Toronto compared to patients in Boston were taller, heavier and had better mean survival by 9 years (30 years in Toronto compared to 21 years in Boston). Since that time it became mandatory to use high fat, high calorie diet in the management of CF patients. Levy[11] studied 19 CF patients with enteral and parenteral feeding after which 10 of these patients died after nutritional intervention. He found that factors that predicted the mortality of these patients using multivariant analysis were: average heart rate, the presence or absence of pseudomonas cepacia, PaCO2, and the patient age at the time of intervention. When he applied this equation to another 10 patents with nutritional rehabilitation, he was able to correctly predict the survival of 8 of these patients. He concluded that early nutritional rehabilitation should be started before sever malnutrition status appears and long before development of severe lung infection. Kraemer[16] studied 117 CF patients and divided them in to 3 groups in relation to the type of organ involvement: (a) predominantly pulmonary but no gastro-intestinal (GIT) symptoms, (b) pulmonary and GIT symptoms, (c) GIT symptoms only. He found that relative under Wt. is most pronounced in patients with predominantly pulmonary symptoms, and the degree of under Wt closely correlated inversely with survival. In this report we present the implication of gastrostomy tube feeding of Cystic fibrosis (CF) population in Saudi Arabia and its effect on mortality.

METHODS

The charts of all CF patients referred to the CF clinic during the period from November 1994 to October 1998 were reviewed. Cystic Fibrosis was diagnosed on typical clinical picture and high sweat chloride test in two consecutive tests >60 mmol/L by the guantitative method (Wescor, USA). The following definitions have been applied to our results:

Calculated weight (CWT)[6]: Express actual weight as a percentage of ideal body Wt (IBW) = Actual Wt X100 / IBW for Ht.

Calculated height (CHT)[6]: Actual Ht X 100 / 50th percentile Ht for age.

Nutritional failure: Wt for height index below 85% of ideal Wt/ standard Ht, loss of Wt for > 2 month and or plateau in Wt gain for 2-3 month[6].

Z score: It is the standard deviation of Wt and or Ht from the mean of a reference population (Table 1), e.g. if a patient Wt for Ht is at 97th percentile, Z score will be (+1.9), but if another patient parameter at 17th percentile, his Z score will be (–1.0).

Tha data were analyzed on IBM, PL300, computer using JMP program version 3.2 from SAS institute. All variables with normal distribution, mean, standard deviation (SD) and median were calculated using student t-test, other wise a non-parametric test is used Wilcoxon test. For categorical variables, Chi-square of First exact test was used. Uni-variant analysis was performed in all variables. Results were presented at a level of significance of p= <0.05. All values were expressed in mean ± SD.
Epi Info version 6 (CDC, 1994) was used to calculate Z-score, namely Wt for Ht Z-score, Ht for age Z-score, and Wt for height percentile and Ht for age percentile. Mean and standard deviation was calculated for all scores. Each score was compared with the previous score using paired t-test. P value of less than 0.05 was considered significant. We used Bonferroni correction because of multiple comparisons. JMP (version 3.2, 4SAS institute Inc. Carey, NC, USA.) was used to produce the statistical analysis.

Patient management: All confirmed CF patients had their Wt and Ht measured in the first visit and each follow up visits thereafter, which are usually every 2-4 month. All patient with signs and symptoms of pancreatic insufficiency as diarrhea or positive fat in the stool are started on pancreatic enzymes according to CFF recommendation[6] and fat-soluble vitamins (A, D, E and K). Nutritional management and intervention are done by a specialized nutritionist according to CFF recommendations[6]. Oral supplement with elemental formula like Soya base formula in patients less than 1 year of age or Pediasure (Lactose, gluten free, fortified formula with 30 calories/ Ounce) in patients more than 1 year of age. Nasogastric (NGT) feeding or gastrostomy (GT) feeding were started 2 month after failure of oral supplement to improve weight gain.

RESULTS

Ninety-six CF patients were diagnosed on typical clinical picture and sweat chloride test >60 mmol/L during the period November 1992 - November 1998. Eighty-one (84%) CF patients are a live, Fifteen (16%) died. Fifty (52%) were males and 46 (48%) were females. Age at diagnosis 2.9 ± 3.5 years. The mean follow up period was 3.24 ± 2.8 years with a range of 0.01- 13.8 years. Mean Wt at diagnosis 9.5 ± 7 kilogram (kg), a range of 2.4-36 kg. Calculated Wt/Ht 82 ± 19%, a range of 40 ± 162%. Sixty five percent of patients were in the mild to moderate malnutrition stage (<90th percentile) (Table 1), and 35% in the normal level for Wt/Ht. Calculated Ht/age a mean of 91 ± 12 centimeter (cm), 63% are in the mild to moderate stunted growth (Table 1). Albumin levels a mean of 40 ± 0.5 (N= 35-50g/L). There were no significant difference in all variables between males and females. Wt/Ht Z score (Table 2) has shown improvement in the first 6 month from (-1.7 ± 0.16) to (-0.77 ± 2) (P value= 0.0001), but developed a pleatu level thereafter at 12,18 and 24 month with Z score (-0.86 ± 1.01), (-0.89 ± 1.01) and (-0.93 ± 1.01) respectively (P value= > 0.05) (Table 2). Ht/ age Z score has shown no significant improvement in the first 12-

month, but better response at 18 and 24 month. GT feeding was established in 8 patients, 5 were females and 3 were males. Age when GT inserted was 5.5 ± 2.8 years, a range of 2.6-11.5 years. 2 were still a live at the time of the study and 6 died after GT intervention. Duration of GT feeding was 13 ± 11 months, a range of 1.4-38 months. The arterial blood gas of patient with GT feeding at diagnosis showed: PH 7.4 ± 0.07 (Normal 7.35- 7.45), Partial pressure of carbon dioxide (PaCO2) 5.8 ± 1 k Pascal (N= 4.2-6.0), Partial pressure of oxygen (PaO2) 6.9 ± 2.5 k Pascal (N= 9.8-14.2), Bicarbonate level (HCO3) 30 ± 10 mmol/L (N= 21.0-25.0), and base excess (BE) 9.8 ± 7.2 mmol/L (Normal -2.0- 2.0), a picture of mild compensated respiratory acidosis and metabolic alkalosis with mild hypoxemia. The weights of all patients with GT improved minimally in the first 6 months, but remained the same for at least 2 years thereafter (Fig 1). The height picked up after 6 month of rehabilitation and continued minimal improvement until 2 years thereafter (Fig 2). Comparison of all CF patients who are a live at the time of the study and patients with GT feeding have shown factors that contributed to early mortality of GT patients were: mucoid pseudomonas colonization at diagnosis (p 0.03) and development of early resistance to Genta (p 0.04) (Table 3).

Table 1. Cystic fibrosis growth parameters in saudi population

VARIABLE	NUMBER	%	PERCENTILE	STATUS
CWT	33	34	< 75%	Severe malnutrition
	7	7	75- 79%	Moderate malnutrition
	11	11	80- 84%	Mild malnutrition
	11	11	85- 89%	Under weight
	30	31	90- 110%	Normal weight
	4	4	> 110%	
CHT	19	20	< 85%	Severe stunted
	8	8	85-89%	Moderately stunted
	24	25	90- 94%	Mildly stunted
	36	37	95- 100%	Normal

CWT= calculated weight for age
CHT= calculated height for age

Table 2. Weight for height z score during 2 years follow up

PERIOD		MEAN SCORE	NUMBER	P= VALUE
WHZ at Diagnosis		-1.7 ± 0.16	74	
WHZ at 6 month		-0.77 ± 2	74	0.0001
WHZ	12 M	-0.86 ± 1.01	58	
Z score	12 M and 6 M			0.918
WHZ	18 M	-0.89 ± 1.01	48	
Z score	18 M and 12 M			0.588
WHZ	24 M	-0.93 ± 1.01	46	
Z score	24 M and 18 M			0.703

WHZ= weight for height Z score
DX= Diagnosis, M= month

Table 3. Comparison of patients with GT and the general CF population

CHARACTER	CF PATIENTS (88)	GT PATIENTS (8)	P VALUE
Sex Female Male	46 42	5 3	0.8
Status Alive Died	79 9	2 6	0.0001
Age DX (a)	2.9 ± 3.5	4.1 ± 1.7	0.94
Age FU (b)	6 ± 4	6.7 ± 2.7	0.6
WT/ HT (c)	82 ± 19	72 ± 13	0.17
HT/Age (d)	91 ± 12	94 ± 5	0.4
R Ampicillin (e)	73	7	0.23
Pseud +ve (f)	55	6	0.2
Mucoid Pseud +ve (g)	25	6	0.03
R Gentamycin (h)	19	3	0.04

(a)= Age at diagnosis
(b)= Age at follow up
(c)= Weight for height percentile
(d)= Height for age percentile
(e)= Pseudomonas resistance to Ampicillin
(f)= Positive culture for pseudomonas aeruginosa
(g)= Positive culture for mucoid pseudomonas
(h)= Pseudomonas resistance to Ampicillin

Figure 1. WT= Weight
GT= Gastrostomy tube feeding

Figure 2. HT= Height
GT= gastrostomy

DISCUSSION

Many studies have shown that parenteral nutritional interventions either through NGT or GT have markedly improved survival. Levy et al[15], studied 14 patients on gastrostomy feeding for 1 year and found that supplemental feeding resulted in increase of Wt as % of standard by 2% in the treated group but declined by 3% in the control group. Forced vital capacity (FVC) didn't change in the treatment group, but

declined by 12% in the control group. Forced expiratory volume in one second (FEV1) declined by 13% in the control group, but remained un-changed in the treatment group (p value <0.01). He also noted that in the treatment group, there was a marked increase in the ability to participate in the activity of daily living. Shephard et al[13], followed 10 patients with NGT feeding for 2 years, and found that long term nutritional rehabilitation resulted in a catch up weight gain and sustained improvement in linear growth, fewer pulmonary infections/ year, a significant reversal of the deteriorating lung function. Protein synthesis exceeded protein breakdown by one-month supplementation. The mean change in 2 Z scores for Wt and Ht were significantly grater in treatment group compared to control group as no catch up growth was noted in the latter. Boland[17] studied 10 patients with nocturnal jejunostomy feeding for 10-36 months and found that all patients gained weight and had improved activity, with stabilization of their pulmonary function test and improvement in quality of life for both patents and their families. In our study we have shown that more than 85% of our patient were in the mild to moderate level of malnutrition at presentation, but Z score improved the first 6-12 month after starting pancreatic enzymes and vitamins and initial oral nutritional rehabilitation, but gradually plateau or progressively decreased due to progressive lung disease. Only 8 patients had GT insertion for feeding due to refusal of most parents and patients for this type of intervention for fear of social and school embarrassment and to maintain the patient's self integrity and body image. GT feeding did not improve the survival of our CF patients due to delay in GT insertion until development of severe mal-nutrition stage and actually they died shortly thereafter due to progressive malnutrition and deterioration of lung disease. Other factors that may need to be considered for early mortality is multi resistant bacteria (Table 3), and poor compliance to treatment and chest physiotherapy as most parents have other healthy siblings to take care of and ignore the sick child.

CONCLUSION

Early nutritional rehabilitation is necessary to improve survival in CF patients before deterioration of lung disease and development of severe malnutrition. Efforts should be encouraged to improve the knowledge of the usefulness of NGT and GT feeding and to disseminate the knowledge on the sequela of severe malnutrition.

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