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To assess the need of palliative care in children with life limiting diseases using Paediatric Palliative Screening Scale (PaPaS Scale)

Authors
Dr Rakesh Sharma[1], Dr Gaurav Sharma[2], Dr Deepak Sharma[3], Dr Pancham Kumar[4], Dr Jyotsna Sharma[5]
[1]Professor, Deptt. of Paediatrics, Indra Gandhi Govt. Medical College, Shimla, H.P.
[2]Junior Resident Deptt. of Paediatrics, Indra Gandhi Govt. Medical College, Shimla, H.P.
[3]Medical student, MMU Solan
[4]Assistant Professor, Deptt. of Paediatrics, IGMC, Shimla
[5]Senior Resident, Indra Gandhi Govt. Medical College, Shimla, H.P.
Corresponding author
Dr. Gaurav Sharma
IGMC Shimla 171001, India
Email: *Gaurav.rpgmctanda@gmail.com*, Phone number- +91 9418885096

Abstract
This is the first ever study in northern India to assess the need of palliative care in a tertiary care setting comprising of 102 children during one year period interval in department of paediatrics at IGMC Shimla, Himachal Pradesh. Modified Paediatric Palliative Screening Scale (Modified PaPaS Scale) a performa based oral questionnaire was used to assess the need of palliative care in the study groups. It was found that patients with severe cerebral palsy, HIV, DMD and MPS required aggressive palliative care and needs to be initiated as early as possible. To summarise, our study supports creation of a hospice based palliative care facility to all patients with chronic and life limiting disease.

Introduction

The term "palliative" is derived from the Latin word *pallium* meaning a cloak (conceal or alleviate). Palliative care aims to cloak the patient's symptoms and provide comfort even when treatments aimed at cure are no longer possible. Pediatric palliative care is an interdisciplinary collaboration that seeks to improve the quality of life of all children with life-threatening conditions, as well as their families(1). It focuses on prevention and relief of suffering, regardless of the stage of disease, and comprehensively addresses the physical, psychosocial, or spiritual needs of the child and family(2-4). A number of scales are designed to improve recognition of the need for palliative care (5-6) e.g. Edmonton Symptom Assessment Scale (ESAS), Pediatric Palliative Screening Scale (PaPaS Scale) etc.

Material and Methods

Prospective study was carried out in the department of paediatrics IGMC Shimla, Himachal Pradesh. Assessment of the palliative care needs was done in both inpatient and outpatient hospital setting. This study comprised of 102 children who presented during the study period from 1st June 2016 to 31stmay 2017 and satisfied the inclusion and exclusion criteria. The Inclusion Criteria for the study included Children from 1-18 yrs of age with life limiting diseases. Life limiting diseases was defined in the study as disease which falls in the below mentioned four groups.(7-8)

Group 1	life threatening conditions for which curative treatment is feasible but can fail. This group included children with Cancers, heart defects, irreversible organ failures
Group 2	Conditions where premature death is inevitable. Treatment may aim at prolonging life and allowing normal activities. This group included children with cystic fibrosis, Duchenne muscular dystrophy
Group 3	Progressive conditions without curative options. Exclusively palliative treatment may extend over many years. This group included children with metabolic disorders, neuromuscular diseases
Group 4	Irreversible but non progressive conditions causing severe disabilities leading to susceptibility to health complications and likelihood of premature death. Children with severe cerebral palsy were included in this group

Neonates and Infants were excluded from the study.After distribution of the children in the above mentioned groups written informed consent was obtained from the parents/guardian for the participation in the study.

Modified Paediatric Palliative Screening Scale (Modified PaPaS Scale) which is a performa based oral questionnaire on following domains:- Domain 1 - Expected life expectancy. Domain 2- Expected outcome of current treatment and burden of this treatment. Domain 3-Performance status. Domain 4- Symptom and problem burden. Domain 5- Preferences of patient, family and health professional.The total score at the end of questionnaire was obtained by adding the scores in the five domains.

The need of palliative care in the group was assessed based on the following score: When the total score was less 15, these children were considered having no palliative care needs. The children with total score more than and equal to 15 and less than 25, were considered to prepare for start of palliative care.For children with Score equal to or greater than 25, need palliative care and the care to be started immediately.

Results

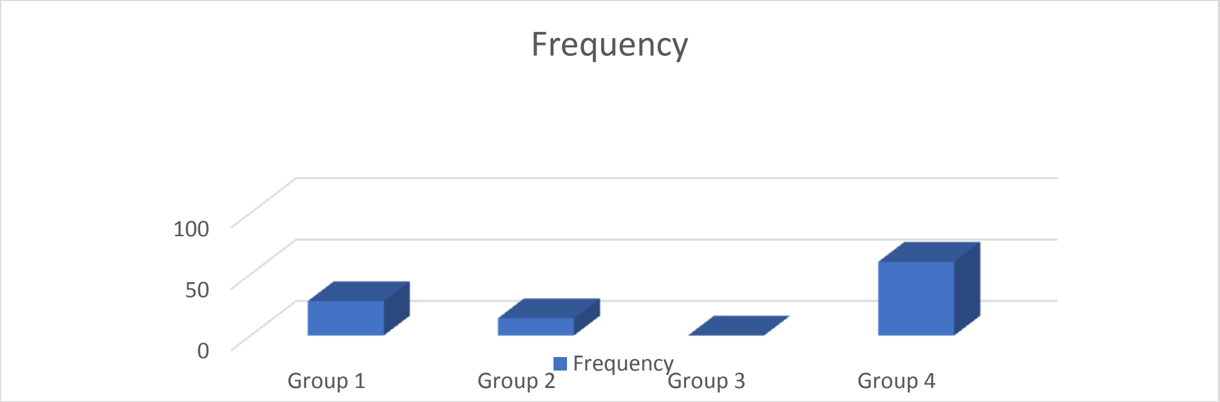
Out of the 102 children, males predominated the study population being 59 in number as compared to 43 females. Maximum number of children were in age group of 1-5 years (n=65) and least being in 10-15 years (n=7).

1. Group 1 included a total of 28 cases. Out of the 28 cases, 25 were cases of haematological malignancy and 3 cases of complex congenital heart disease.
2. Group 2 included a total of 14 children out of which 7 were cases of DMD, 4 were cases of mucopolysaccharidosis and 3 cases of HIV.
3. Group 3 No child with metabolic disorder and neuromuscular disease was enrolled in this group.
4. Group 4 60 children of severe cerebral palsy were enrolled in this group.

Chart 1: frequency distribution of groups

Group 1

	Haematological malignancy	Heart defects	Total
Number of cases	25	3	28
Mean age	5.8 years+-2.84	4 years+-2	5.6years+-2.79
Mean total score	12.72.+-6.2	29.66+-1.52	14.7+-7.99



Out of 25 children with haematological malignancy (ALL 2) had score greater than 25 while the rest had score less than 25.All the children having complex congenital heart disease (2 with TOF with PS and 1 with VSD with eisenmenger syndrome) had score greater than 25.

Group 2

	DMD	MPS	HIV	TOTAL
Number of cases	7	4	3	14
Mean age	6.8 years+-2.19	6.25 years+-2.87	9 years+-5.19	7.14+-3.05
Mean total score	25.14+-5.5	22.75+-3.59	33+-7.9.	26.14+-6.45

Out of 7 children with DMD 4 children had score greater than 25 and 3 child in the age group had a score of 15-24. Among children with MPS 2 children had score greater than 25 and 2 had score of 15-24. 2 children of HIV had score of greater than 25 while 1 child had score between 15-24.

Group 4

	CEREBRAL PALSY
Number of cases	60
Mean age	4.21 years
Mean total score	18.2

50 children with severe cerebral palsy had score 15-24 while 10 children had score greater than 25.

Discussion

This study is first of its kind in northern part of India aiming to enhance quality of life for seriously ill children and ease the pain and suffering of their parents by improving and encouraging early access to paediatric palliative care.

Our study was to assess the need of paediatric palliative in children with chronic debilitating diseases In our study around 61% patients were in the age group of 1-5 years, 33% patients were in the age group of 5-10 years and 7% in the age group of 10-15 years. In the study by Moya et al, 33% patients were in the age group of 1-7 years, 24% patients were in the age group of 8-12 years and 12% were in the age group of 13-17 years.(9) They also included newborns and infants, which were excluded in our study and their study was a large scale study, moreover the age group taken was different than our study explaining the difference in the results.

In our study we divided children with chronic debilitating diseases into four groups, out of which 24.5% children had haematological disease, 2.94% cases had Complex congenital heart disease and 6.86% cases were of DMD. In the study by Moyà et al children there were 22% children with haematological diseases, 5% cases of complex congenital heart disease and 7% cases of DMD(9). This distribution is coinciding with our study.

Cases of cererbral palsy in our study was 58.8%, whereas study done by Moyà et al the included 34% of children with CNS abnormality.

This difference in number of cases may be accounted by the demographic variations in prevalence of disease in particular cerebral palsy.

In another study Monterosso et al instead categorised children into two groups. He divided children into cancer (14.7%) and non cancer group (85.3%) in his study.(10). In our study children with cancer were 24% and non cancer children were 76% which is almost comparable.

We assessed the need of paediatric palliative care using modified paediatric palliative care screening scale. The objective of the PaPaS Scale; (Annexure 1) is to improve awareness of Paediatric palliative care and its potential benefit for severely ill children and their families. The endpoint of the PaPaS Scale is the outcome of a complex process of decision- making. Formulated in the year 2013 its further validity was checked in a study by Eva Bergstraesser et al and Michaela Paul et al in the year 2014.(11) Most studies in the past which were conducted on a large scale took mortality in disease to assess the need of paediatric palliative care in children, this screening scale however takes morbidity into account for assessment of need.

Conclusion

Our study is different from all other studies published because we are the first one to study need of palliative care in a tertiary care setting and also the first one in northern India. We found that most patients with severe cerebral palsy, HIV, DMD and MPS will require palliative care and this should be started immediately as early as possible. This calls for creation of a hospice and palliative care facility in all hospitals and all patients with chronic and life limiting disease should be assessed on diagnosis for the need of palliative care.

Declaration

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Declaration- none

Ethical issues- permission taken from ethical committee IGMC Shimla H.P.

References

1. Field MJ, Behrman RE. Institute of Medicine (U.S.). Committee on Palliative and End-of-Life Care for. When children die: improving palliative and end-of-life care for children and their families, National Academy Press, Washington DC 2003.
2. World Health Organization. WHO definition of palliative care. http://www.who.int/cancer/palliative/definition/en/ (Accessed on March 15, 2008).
3. ACT and the Royal College of Paediatrics and Child Health (RCPCH). http://www.act.org.uk/content/view/77/127/ (Accessed on March 15, 2008).
4. Fraser J, Harris N, Berringer AJ, et al. Advanced care planning in children with life-limiting conditions - the Wishes Document. Arch Dis Child 2010; 95:79.
5. Glare PA, Eychmueller S, McMahon P: Diagnostic accuracy of the palliative prognostic score in hospitalized patients with advanced cancer. J Clin Oncology. 2004, 22 (23): 4823-4828. 10.1200/JCO.2004.12.056.
6. Maltoni M, Caraceni A, Brunelli C, Broeckaert B, Christakis N, Eychmueller S, Glare P, Nabal M, Vigano A, Larkin P: Prognostic factors in advanced cancer patients: evidence-based clinical recommendations–a study by the Steering Committee of the European Association for Palliative Care. J Clin Oncology.
7. Fraser J, Harris N, Berringer AJ, et al. Advanced care planning in children with life-limiting conditions - the Wishes Document. Arch Dis Child 2010; 95:79.

8. Himelstein BP, Hilden JM, Boldt AM, Weissman D. Pediatric palliative care. N Engl J Med 2004; 350:1752.
9. Juan Carlos Irola Moya et al, and Miriam Garro Morales et al Pediatric Palliative Care, Costa Rica's Experience American Journal of Hospice& Palliative Medicine ® 27(7) 456-464The Author(s) 2010 Reprints and permission: sagepub.com/journalsPermissions.nav DOI: 10.1177/1049909110361471 http://ajhpm.sagepub.com
10. Leanne Monterosso et al. Supportive and palliative care needs of families of children with life-threatening illnesses in Western Australia: evidence to guide the development of a palliative care service, Palliative Medicine 2007; 21: 689–696.
11. Eva Bergstraesser et al , Michaela Paul et al , Kaspar Rufibach et al , Richard D Hain et al and Leonhard Held et al, The Paediatric Palliative Screening Scale: Further validity testing, Palliative Medicine2014, Vol. 28(6) 530–533, The Author(s) 2013.

Abbreviations

PaPaS Scale- Paediatric Palliative Screening Scale
DMD- Duchenne muscular dystrophy
MPS- mucopolysaccharidosis
HIV- human immunodeficiency virus
ALL – acute lymphoblastic leukaemia
TOF with PS-tetralogy of fallotwith pulmonary stenosis
VSD- ventricular septal defect

Annexure 1

Domain and Item number	Item	Characteristic	Score (preliminary)
Domain 1	**Estimated life expectancy**		
1.1	Estimated life expectancy	> 2 years	0 □
		> 1 but < 2 years	1 □
		3 months to 1 year	2 □
		< 3 months	3 □
1.2	"Would you be surprised if this patient were still alive in 6 months time?"	Yes	3 □
		No	0 □
Domain 2	**Expected outcome of current treatment directed at the disease and burden of this treatment**		
2.1	Expected outcome of treatment directed at the disease	There are no treatments currently that can cure the disease or prolong life.	4 □
		Current treatment patient is receiving or will be receiving may cure the illness.	0 □
2.2	Burden of treatments	Treatments carry a high level of burden (many side effects).	2 □
		Treatments carry no or minimal burden (side effects) or no treatment is envisioned.	0 □

Domain and Item number	Item	Characteristic	Score (preliminary)
Domain 3	**Performance status**		
3.1	Current performance status (in comparison with the child's own baseline)	Moderate to severe restriction of play (no active play, requires assistance for quiet play) 0-40% of normal range.	3 □
		Mild to moderate restriction of play (able to engage in some active play; requires assistance) 50-70% of normal range.	1 □
		Normal range of play (able to carry on usual play activities) 80-100% of normal range.	0 □
3.2	Rate of decline of performance status	Overall, performance has decreased by half over the last 4 weeks.	2 □
		Overall, performance has decreased by about a third over the last 4 weeks.	1 □
		Overall performance has not deteriorated over the last 4 weeks.	0 □
Domain 4	**Symptom and problem burden**		
4.1	Number of symptoms	Patient has 3 or more symptoms (e.g. pain, weight loss, fatigue, dyspnoea, nausea & vomiting, depression, anxiety)	4 □
		Patient has 2 symptoms	3 □
		Patient has 1 symptom	2 □
		Patient is asymptomatic	0 □
4.2	Symptom intensity As perceived by the parents	Any symptom is severe (equivalent to >6 out of 10)	3 □
		Any symptom is moderate (equivalent to 4–6 out of 10)	2 □
		Any symptom is mild (equivalent to 3 or less out of 10)	1 □
		Symptoms are absent	0 □
4.3	Psychological distress of patient	Significant	2 □
		Mild to moderate	1 □
		Absent	0 □
4.4	Psychological distress of parent(s)	Significant	2 □
		Mild to moderate	1 □
		Absent	0 □

Domain and Item number	**Item**	**Characteristic**	**Score (preliminary)**
4.5	Psychological distress of siblings	Significant	2 ☐
		Mild to moderate	1 ☐
		Absent	0 ☐
Domain 5	**Preferences of patient, family and health professional**		
5.1	Request by patient and family	Patient specifically requests a palliative care approach.	4 ☐ Yes 0 ☐ No
		Family specifically requests a palliative care approach.	4 ☐ Yes 0 ☐ No
5.2	Preference of health professional	You feel that this patient would definitely benefit from a palliative care approach.	4 ☐ Yes 0 ☐ No
		Total score:	