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Editorial

It is an honor to edit this edition of IJGDBP where two very diverse yet inseparable groups of medical science, growth and development come together.

The theme of this issue is Screening and Diagnosis. In the realm of developmental and behavioral subjects, the issue of objectivity remains a moot factor where it is considered subjective by many experts. It is therefore, exhilarating to observe this positive metamorphosis (or shall we say development), which has a propensity to propel this important branch of pediatric practice to make it safe for children, especially the most vulnerable of them, the children with special needs.

Fundamental human activities like eating, sleeping or shoe lacing can be objectively measured. Only if we can accurately measure them as much as possible, we can do justice in mending them. This, as a concept, has shaken even the most scientific of its minds in the past. It is only in the last few decades developmental pediatricians are beginning to grapple with this concept doing justice to this issue.

We have interesting articles on screens and diagnoses regarding eating, drawing or tooth brushing in children that a pediatrician can use in his or her clinic when (s)he sees a child facing the difficulty. Dr MP Mohanta, Dr Sunil Godbole and Mr Jewel Chakraborty have done a fabulous job here.

Dr Nandita Chattopadhyay brings a somber perspective to the reality of global scenario where the most fundamentals of human rights are denied to our future citizens in resource deprived settings.

Anjan Bhattacharya
Editor of this Issue

About the Editor

Dr Anjan Bhattacharya is a developmental pediatrician and he was instrumental in setting up of the Child Development Centre at Apollo Gleneagles Hospital, Kolkata. The impact of this effort can be seen in the children with EBD (Educational/Emotional, Behavioral and Developmental) problems, which constitute 1 in 5 to 1 in 6 children throughout the globe.

This was recently recognized internationally by the World Health Organization whereupon he participated in the collaborative pioneering research of WHO on core standard setting of ICF in children, using cerebral palsy as its pioneering model. He is currently coordinating the multicentric international validation research of this tool for our WHO regions in collaboration of Universities from India, Pakistan and Sri Lanka under the University of British Columbia, Vancouver, Canada.

Dr Anjan Bhattacharya has worked in Newham University Hospital and Primary Care Trust, London as a developmental pediatrician following his DCH and MRCP from Royal College of Physicians of London. He was awarded Purbanchal Pioneer Award by East Zone Academy of Pediatrics and Bharat Jyoti Award by India International Friendship Society. He is currently one of the first 15 National Honorary Fellows in Childhood Disability and Early Intervention conferred by Childhood Disability Group of Indian Academy of Pediatrics.
Introduction
The significance of children’s drawing ability has been extensively explored since the late nineteenth century. They are thought to provide indications of visual-motor development, levels of cognitive functioning, intellectual maturity, projections of personality and self-concept, and assessments of emotional state and disturbances. Drawing is an activity that children tend to enjoy. They willingly produce spontaneous scribbles and drawings from a young age (Koppitz, 1968), making it an attractive method to use in clinical settings.

One of the earliest drawing tests was the Draw-a-Man test devised by Florence Goodenough in 1926 to assess children’s creativity, mental age and visual-motor-intellectual maturity by coding features of their drawing of a man (Goodenough, 1950).

Drawings are believed to be a universal communication tool and an example of non-verbal measurements (Golomb, 2000). Many authors reason that drawings can be analyzed to find underlying messages, such as feelings or indications of interpersonal relationships (Koppitz, 1968; Machover, 1980; Naglieri, et al, 1991). Drawings of human figure are universally one of the most popular subjects in children’s drawings (Golomb, 2000).

Over the years, many different versions of how to study human drawings have emerged. Starting from Goodenough’s Draw-a-Man test (1949) till most recent version, Draw-A-Person: Screening Procedure for Emotional Disturbance (DAP: SPED), many adaptations and modifications are developed with emphasis on different applications.

The non-verbal nature of Draw-A-Person (DAP) test minimizes the effects of formal learning and language barriers and is therefore more suitable for cross-cultural use than static verbal measures (Foxcroft and Roodt, 2004).

The applications of DAP are multifold. In view of pediatric office practice, following applications are noteworthy.

DAP as a cognitive measure
The DAP in its earliest form was developed in 1926 by Goodenough, who realized that the increased sophistication in children’s drawings was related to cognitive ability rather than to chronological age (Goodenough, 1950). Developmental and intellectual levels are reflected in the human figure drawings of children and have been standardized as an intelligence test for children between the ages of three and thirteen years by Goodenough (1950).

Test administration
The child is advised to sit comfortably at a table and given a standard size (A4) plain paper with crayons or pencil. Simple directions are given, such as “I want you to make a picture of a person. Make the very best picture that you can. Take your time and work very carefully. Try very hard and see what a good picture you
can make.” There should not be any time limit. Usually 10 minutes will suffice with young children. No instruction on age, sex, location, time period, or medical condition should be given. The DAP and its revisions come with standardized instructions and checklists to score each feature of the drawing, scoring manual and result interpretation. It requires training, which is not very difficult for one who has education and experience of dealing with children.

In 1963, Harris revised the test as the Goodenough-Harris Drawing Test (GHDT) by adding two additional drawings of a woman and the self. In 1988 Naglieri further updated the test as DAP: A Quantitative Scoring System (DAP: QSS). Norms were established on children between ages six to seventeen years from different cultural and socio-economic status.

The rationale behind the DAP as a cognitive measure relates to the belief that developmental changes in children’s drawings are concurrent with cognitive development (Golomb, 2002). Following developmental stages of drawing were observed.

1. Uncontrolled / basic scribble 13 months approximately
2. Controlled scribble 2 to 4 years
3. Pre-schematic stage 4 to 5 years
4. Symbolic stage 5 to 6 years
5. Schematic stage 6 to 9 years
6. Emergence of realism 9 to 11 years

These developmental stages are approximate and variation does occur between children. But still this knowledge of developmental peculiarities in drawing is important when using drawing for assessment purposes. The cognitive and motor development of children thus affects their human figure drawing. Even certain inclusions, omissions might be characteristic of a certain developmental stage. Kellogg (1979) also observed that making marks on the paper is a stimulus for brain development in areas such as reading and letter formation. This too forms the basis of the DAP as a measure of cognitive development.

That Draw-A-Person test became extremely popular can be explained by its advantages.

(i) Satisfactory correlation with other IQ tests
(ii) The ease of administration
(iii) The pleasure of the subjects and
(iv) Its use in case of impossibility of usage of some verbal test.

In India, Dr Pramila Phatak developed Indian norms for Draw-A-Man test (Mukherjee and Nair, 2011).

Using children’s drawings to assess cognitive ability has limitations due to high levels of misclassification, i.e. false positives and false negatives (Willcock, et al, 2011). There are also other limitations to the cognitive interpretations of DAP as the drawings become unpredictable in pubertal age, disabled children, mentally challenged children, attention deficit hyperactivity disorder, autism spectrum disorder and obsessional disordered patients.

The test does not claim to be an IQ test in itself but it can provide a good representation of the level of the intellectual functions of the subject. In busy pediatric OPD practice the DAP could be a quick, handy screening tool to approximately judge the cognitive capacities of a child.

**DAP as projective test**

The traditional method of assessing whether a child has an emotional or behavioral problem is by administering behavior checklists to the parents, teachers and caregivers. As these checklists are completed by individuals other than the child themselves, any conclusions that are made based on such measures cannot be conclusive, as some facets of behavior may not be identifiable by the parent, teacher or
Caregiver. To obtain diagnostic information directly from a child, several issues may be encountered such as language boundaries, limited understanding of emotion and inability to verbalize feelings, or even responding in an acquiescent way. To counteract such issues, drawing tests which involve the objective interpretation of emotional profile of child were developed (Miranda Cruso, 2013). This type of testing, also known as projective testing is designed to let a person respond to ambiguous stimuli, presumably revealing hidden emotions and internal conflicts projected by the person into the test. This is sometimes contrasted with a so-called "objective or self-report test" in which responses are analyzed according to a presumed concept and are limited to the content of the test (Machover, 1980).

In 1949, Karen Machover developed the first measure of figure drawing as a personality assessment with the Draw-A-Person Test. Children's drawings are indicators of emotions, self-esteem and social competence, as well as other aspects of personality (Di Leo, 1973) and this information can be used in counseling. Drawings allow children to experience and express rather than verbalize feelings. Machover's projective application of Draw-A-Person Test studies human figure drawing of a child subjectively rather than quantitatively. Various aspects such as size of the head, placement of the arms and even trivial things such as if teeth were drawn or not are thought to reveal a range of personality traits. The personality traits can be anything from aggressiveness, to relationships with their parents, to introversion and extroversion.

Later Koppitz developed a quantitative measure of assessment that has a list of emotional indicators including size of figures, omission of body parts, and some "special features". The total number of the indicators is simply added up to provide a number that represents the likeliness of disturbance (Koppitz, 1968). The most recent and psychometrically advanced human figure drawing test for assessing emotional problems is the Draw-A-Person: Screening Procedure for Emotional Disturbance (DAP: SPED), developed by Naglieri, et al, (1991). The test was developed as a modification of existing objective approaches to scoring human figure drawings (HFD), based on physically measured structural items and inclusions or omissions of particular content items.

**Test administration**

DAP: SPED also follows similar instructions as those of basic Goodenough Draw-A-Man test; but it differs on few points. Three instead of one standard size (A4) papers are given. On first paper the child is instructed to 'Draw-A-Man'. Once completed, the child is advised to make the best picture he/she can and remind him/her, please, to draw a whole man, not just the head and shoulders. The scoring is done strictly as per the norms outlined by Naglieri, et al in the DAP: SPED manual. The raw scores of all the drawings are considered together and interpreted. As compared to Goodenough's Draw-A-Man test this projective test requires lot of training and deep insight of psychology. Obviously this becomes a limitation for its use as a screening tool in pediatric office practice.

**Limitations**

There are some limitations to DAP as a projective tool. For example, children with drawing talent draw the human figure with more details, sometimes shading, tend to use eraser more often till they find drawing artistically better. But this can be wrongly interpreted as insecurity or conflicts (Thomas,1990). Every child's developmental stage is different; so as their drawing skills. If the developmental stage is not considered while interpreting DAP the
results may be erroneous. Interpretation of DAP of mentally and/or physically challenged children should be done cautiously.

**DAP as a rapport building tool**

Di Leo (1983) states that drawings are one means of establishing a rapid, easy, pleasant rapport with the child. Children who are shy, quiet, impulsive, have speech and language difficulties or speak a different language from the counselor usually respond well to drawing activities. Drawing a human figure is a simple and direct way of communicating because it deals with what he does rather than with what he says. For this reason, it is more specifically described as an expressive medium. But there are some limitations too. Many small children willingly draw pictures at the request of adults but as children reach adolescence they are reluctant to draw (Rubin, 2005).

**Conclusion**

Children’s draw-a-man test can be used in many ways. Since the Draw-A-Person Test (DAP) is a brief and easily administered test which requires little material, appeals to most children, and yields both an estimate of mental age and considerable projective data, it is extensively useful in psychological evaluation of children. Through the use of such activity, pediatricians can build rapport with children, observe and listen to their thoughts and feelings, discuss important issues, design interventions as appropriate and refer to counselors, therapists, special educators if necessary.

**Further reading:**

1. Koppitz E M. Psychological Evaluation of Children's Human Figure Drawings, 1st edn. London: Grune & Stratton, 1968.
Importance of Early Markers in Autism

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Keywords: Autism, ASD, Early markers

One of the most challenging neuro-behavioral problems in children today is Autism Spectrum Disorder or ASD. A sharp rise in incidence is being noted, largely due to heightened awareness among parents, teachers and physicians. It is an encouraging fact, now-a-days, that a good number of children are being diagnosed by 3 years of age, whereas, even 5 years back, we would get undiagnosed patients coming in at 5-8 years. But there is no scope for complacency yet, as many children are still not referred in time by the primary physician. Where are we lagging behind?

Autism is a condition whose neuro-biological basis is established but the underlying neuropathology is not yet clear\(^1\). So we depend largely on behavioral presentation and developmental history for a diagnosis. ASD is characterised by A) persistent deficits in social communication and interactions and B) Restricted, repetitive patterns of behavior, interests or activities, as per DSM V. It is also clearly stated that symptoms must be present in early development period, though may not be fully manifest till social demands exceed limited capacities (DSM 5). No clear age of onset has been mentioned. It has been found that a large proportion of children manifest problems between 12-24 months\(^2,3\), some even before 6 months\(^4\). Yet, while parents may note early signs, there is often a significant delay between the first parental concern and definitive diagnosis\(^5\). This is largely because the subtle early symptoms may be missed or attributed to general developmental problems at the first point of contact. The primary physician often tends to reassure the parents with a comment, “he will grow out of it”. A UK based study with 770 families showed that 25% of ASD were reassured to be developing normally at 2 years and finally diagnosed as late as 5.5 years\(^6\). Parents and immediate care-givers are mostly in a state of denial and the mild features are either ignored or considered a temporary aberration. The features attract attention mostly when the child fails to talk even at 3 years or faces problems in communicating in school. Here again, it is the verbal communication or the scholastic performance that is of vital importance to the parents and many of the behavioral issues are ignored. These are the reasons why, even though parents may be aware of developmental problems early in life, there is often a significant delay between parental concern and diagnosis\(^8\), which may be as long as 3-4 years\(^7\).

Mother – The best assessor

The mother being the most vigilant observer of the baby, she is most likely to pick up the
subtlest cues of atypical behavior. It is the ‘maternal instinct' that evokes this perception, and it is equally sharp and vigilant among intelligent educated urban mothers as well as illiterate, rural mothers. Here I cannot refrain from sharing a personal experience, where a tribal, illiterate mother brought her 15 month old son saying in her own tribal dialect, “Shune, kintu bujhe na” which means ‘He listens but does not understand.’ This child was later confirmed as ASD.

How early can she note the difference?
It may very well be in early infancy. So, we only need to pay heed to what is striking the mother.

What is it that she notices in these early months?
The baby may not look into the mother’s eyes or reciprocate her smile. He/she may show inadequate facial expression and lack of proper interest in people around. The baby may be excessively cranky and difficult to console. Inadequate response to vocal communication, inability to follow a point or imitate gestures (waving and clapping) and some atypical sensory-motor behavior or stereotypes may also be noted by the mother.

Knowledge of these early symptoms has prompted a lot of research across the globe on early markers of Autism. The importance of identification of early markers lies in the fact that it will facilitate early diagnosis and early intervention, which will maximise the developmental outcome. It is an accepted fact that earlier the interventions, greater the chances to move towards a more typical developmental trajectory, because of the neuroplasticity, more so if initiated before 2 years of age. Moreover, the onset of secondary behaviors like aggressiveness, restricted rituals, severe problems with socializing etc can be prevented with early intervention. Interventions provided before language and social skills drop off can even prevent such losses.

What are the early markers?
Based on observations in young children at risk of autism, e.g., siblings of ASD children and retrospective analysis of diagnosed children, a good number of early markers have been identified and categorised for early detection.

The early markers may be grouped under the following heads:

Abnormalities of social attention and communication:
Delays, absence or abnormalities in the following activities are considered as “Red Flag”. It is not just the presence or absence of these typical behaviors, but the quality that needs to be monitored. Evidence of these features from first year of life, are the most predictive early signs.

Social attention –
1. Social games like peek-a-boo or ‘tuki’, with proper eye contact and smiles by 8 months
2. Proper eye contact by 8-24 months
3. Turning to name call (number of prompts?) by 8-24 months
4. Social smiling with directed eye contact by 8-24 months
5. Imitation like waving, clapping by 8-24 months

Communication –
1. Use of language – Babbling by 8 months, 1-3 words by 12 months, 5-10 by 18 months and combining 2 words by 24 months.
2. Understand ‘give me’ by 12 months, follow simple command without gestures by 18-24 months
3. Pointing, with eye contact, by 15 months
4. Joint attention: following another’s point and gaze by 12-24 months
5. Social gestures like nodding by 12-24 months
6. Spontaneous pretend play by 15-18 months
7. Parallel play by 24 months

Aberrant behaviors
These are mostly not so useful predictors of ASD, as they may accompany developmental delay as well.5.

They include the following.
1. Sensory behaviors like visual examination, smelling, licking, tactile exploration, over-sensitivity to noise etc and subtle stereotypic behaviors like hand flapping, tip-toeing, shaking etc. may be noted prior to 24 months.

Conclusion
ASD detection is on the rise in our country, though still at a later age. Early Intervention facilities are still very limited, being available in scanty numbers and restricted to only a few cities. More and more children are being diagnosed, but at a later age, when we are unable to bring about an optimum improvement. If we could focus on screening of early markers at a younger age, we might be able to do justice to this huge socio-medical problem of autism. This will call for intense research for an appropriate screening tool, adequate awareness generation among all stake-holders and involvement of a dedicated health care delivery mechanism in conducting stream-lined screening. Though a herculean task, let us come forth to join the global movement in conceptualizing a methodology for early identification of autism at all levels.

Further reading:


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**Members are requested to submit**

*original articles, review articles, case scenarios*

for

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Screening and Diagnosis of Developmental Dyspraxia

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Developmental dyspraxia was first described by A Jean Ayres who pioneered the theory of SI. Ayres stated that children with developmental dyspraxia often have trouble coping with life situations including childhood activities like play, academic learning and social behavior. This disorder therefore has a profound impact on children and their daily life occupations.

In the previous issues of this journal developmental dyspraxia was discussed by one the authors of the present article as a part of the topic as Developmental Coordination Disorder (DCD).

With any diagnosis, comes screening and diagnostic tools. In this issue, we are committed to focusing on these two aspects alone.

Screening
Developmental Coordination Disorder Questionnaire [DCDQ] was developed in Canada for 5 to 15 year olds in the year 2007 [DCDQ'07] based on DSMIV. It is fully available at www.dcdq.ca

It is self-explanatory, easy to apply and fairly accurate. Readers are directed to it for kind perusal. Validations studies in India could make it even more accurate in local context. We are currently interested to work (1) in adopting it for the At-Risk Group i.e. the 3 – 5 year group, (2) editing it as par DSM 5 criteria and (3) validating it.

Diagnosis
Developmental dyspraxia was first identified with a measurement instrument developed by Ayres in 1972, the Southern California Sensory Integration Test (SCSIT) and later the Sensory Integration and Praxis Tests (SIPT) in 1989. Through development of the SCSIT and the SIPT, Ayres and later Mulligan were able to link poor discrimination of tactile, vestibular and proprioceptive input with dyspraxia. This confirmed association between developmental dyspraxia and sensory discrimination contributed to the development of treatment protocols for developmental dyspraxia.

SI (Sensory integration) is one of the frameworks that underpin developmental dyspraxia as a construct. From a SI perspective, it is essential to have knowledge of the three processes of praxis in order to understand developmental dyspraxia. One of these processes namely ideation, motor planning and motor execution are usually implicated when praxis is deficient.

Developmental dyspraxia consists of four types of dyspraxia that were derived from factor and cluster analysis of the SIPT results. Ayres and Mulligan identified the following types of dyspraxia: Visio-dyspraxia, somatodyspraxia, bilateral integration and sequencing deficits and dyspraxia on verbal command.

The SIPT measures visual, tactile, and kinesthetic perception as well as motor performance. It is composed of the following
17 brief tests:
1. Space visualizations
2. Figure-Ground perception
3. Standing/walking balance
4. Design copying
5. Postural praxis
6. Bilateral motor coordination
7. Praxis on verbal command
8. Constructional praxis
9. Postrotary nystagmus
10. Motor accuracy
11. Sequencing praxis
12. Oral praxis
14. Kinesthesia
15. Finger identification
16. Graphesthesia
17. Localisation of tactile stimuli

The SIPT is scored and interpreted through use of computerized scoring where the subject's raw scores are entered into the SIPT scoring programmer and raw scores are converted to standard deviation (SD) scores.

SIPT test results are expressed in SD scores. Scores between -1.0 SD and +1.0 SD are considered in the average range, score of -2 to -1.0 suggest mild difficulties, score of -2.5 to -2 indicates definite dysfunction, -3 to -2.5 indicates severe dysfunction. A score of +1.0 to +2.0 indicates above average functioning and a score of +2.0 to +3.0 indicates advanced functioning. The SIPT computer generated report briefly describes each test and the obtained standard score, has a summary bar graph that shows the major results, lists various scores such as the Standard error of measurement (SEM), SD scores, measurements of lateral function and an audit of test data. It also shows a summary graph comparing the child’s SD scores to the significant cluster group mean scores.

Bar graph (Fig 1) score shows child has low average scores in the area of
1. Figure-Ground Perception
2. Manual Form Perception
3. Kinesthesia
4. Graphesthesia
5. Localisation of Tactile Stimuli
6. Praxis on Verbal Command
7. Design Copying
8. Constructional Praxis

Fig 1. Child has low average scores
9. Oral Praxis
10. Bilateral Motor Coordination
11. Postotary Nystagmus
12. Postural Praxis

Child has definite dysfunction in the area of Standing/Walking Balance, Motor Accuracy and Severe dysfunction in the area of Space visualization.

Six prototypic groups have been identified to describe the child’s condition including dysfunctional, average and superior patterns of sensory integration. These six groups are:

1. Low average bilateral integration and sequencing
2. Low average sensory integration and Praxis
3. Dyspraxia on Verbal Command
4. Generalized SI dysfunction
5. Visuo & Somatodypraxia
6. High average sensory integration and Praxis.

D squared value of the SIPT scores indicates similarity with these six prototypic groups. Small D squared value indicates a close fit and large D squared value indicates poor fit with these six prototypic groups.

D squared value of below mentioned scores indicates the child has low average bilateral integration and sequencing dysfunction.

Further reading:
Introduction

In civilized worlds, “eating” is no more only ingesting food in response to hunger, but it can be a part of socialization also. That’s not all, eating can be easily affected by our emotions. Because eating is associated with pleasurable feelings due to contact of food with the taste buds, people often seek gratification through eating. This often leads to milliards of problems. Srimad Bhagavatam, which was perhaps composed more than 1200 years ago, gives excellent insights to disordered eating as follows.

“tavaj jitendriyo na syad vijitanyendriyah? puman na jayed rasanam´ yavaj jitam´ sarvam´ jite rase”

“Although one may conquer all of the other senses, as long as the tongue (pleasure of food) is not conquered it cannot be said that one has controlled his senses. However, if one is able to control the tongue, then one is understood to be in full control of all the senses”.

Not the adults only can have ‘Eating Disorders’ (EDs), but the children and adolescents can be also easily affected. EDs in children are common. Diagnosing EDs early is important; because untreated they can pose lifelong misery and ill health. EDs can be subtle in their presentations and need careful evaluation to be detected and diagnosed properly. Pediatricians must be aware of various EDs, their presentations, so as to recognize them in their office practice. Classically ‘Eating Disorders’ (EDs) are described in adolescent girls with inappropriate eating behavior due to misperception of their bodyimage.

But there is an increasing recognition that eating disorders can affect boys and younger children too. Also EDs can be there without any body image misperception. Not surprisingly, with increasing westernization, the incidence of EDs is on rise in India. In the pediatric office practice, many parents are concerned with the eating behavior of their children. Though most of these children may be either normal healthy children or suffering from mild feeding disorders, it is often worthwhile to rule out EDs in such children.

Classification of eating disorders

There is a change in the classification of EDs in the new Diagnostic and Statistical Manual of Mental Disorders fifth edition (DSM5), released in 2013.

Anorexia Nervosa (AN) and Bulimia Nervosa (BN) are the two major EDs, in which the patient is preoccupied with body image and fear of weight gain. It was observed that about half of the EDs did not fit into the above diagnoses and belonged to the Diagnosis of

Screening of Eating Disorders in Children

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‘Eating Disorder not otherwise specified’ (EDNOS) in DSMIV. EDNOS was an umbrella term for sub threshold AN, BN, as well Binge eating and EDs without body image disturbances. This ambiguity in DSMIV has led to introduction of a new category of ‘Avoidant and Restrictive Food Intake Disorder’ (ARFID) in DSM5, whose diagnosis does not require disturbed body image. ARFID replaces the DSMIV diagnosis of ‘Feeding disorders of Infancy and Early Childhood’ which was a nonspecific diagnosis and required lack of weight gain to be criteria. The Binge Eating Disorder (BED) is a separate diagnosis in DSM5 now. Also DSM5 has broadened the inclusion criteria of both AN as well as BN. It is hoped that with this better accuracy in diagnosis, it will be easier to choose specific therapy. The DSM5 Classification of Eating Disorders.

1. Anorexia Nervosa (Table1)
   (a) Restrictive Type
   (b) Bingeeating/Purging Type
2. Bulimia Nervosa (Table2)
3. ARFID (Table3)
4. BED
5. Specified Feeding and Eating Disorders
6. Unspecified Feeding and Eating Disorders

Classification of eating disorders as per the International Classification of Diseases of WHO, 10th Revision 1990, (ICD10)5.
1. Anorexia Nervosa
2. Atypical Anorexia Nervosa
3. Bulimia Nervosa
4. Atypical Bulimia Nervosa
5. Overeating associated with other psychological disturbances
6. Vomiting associated with other psychological disturbances

Table 1 : DSM5 Diagnostic Criteria for Anorexia Nervosa

| A. Restriction of energy intake to requirement, leading to a significantly low body weight in the context of age, sex, developmental trajectory and physical health. Significantly low weight is defined as a weight that is less than minimally normal or, for children and adolescents, less than minimally expected. |
| B. Intense fear of gaining weight or of becoming fat, or persistent behavior that interferes with weight gain, even though at a significantly low weight |
| C. Disturbance in the way in which one’s body weight or shape is experienced, undue influence of body weight or shape on selfevaluation, or persistent lack of recognition of the seriousness of the current low body weight. |

Specify whether

Restricting type:
During the last 3 months, the individual has not engaged in recurrent episodes of binge eating or purging behavior (i.e.; self induced vomiting or the misuse of laxatives, diuretics and enemas). This subtype describes in which weight loss is accomplished primarily through dieting, fasting and/or excessive exercise.

Bingeeating/purging type:
During the last 3 months, the individual has engaged in recurrent episodes of being eating or purging behavior (i.e.; self induced vomiting or the misuse of laxatives, diuretics and enemas).

Specify if:

In partial remission:
After full criteria of anorexia nervosa were previously met, Criterion A (low body weight) has not been met for a sustained period, but either Criterion B (intense fear of gaining weight or becoming fat or behavior that interferes with weight gain) or Criterion C (disturbance in self perception of weight and shapes) is still met.

In full remission: After full criteria for anorexia nervosa were previously met, none of the criteria have been met for a sustained period of time.

Level of severity is based on current body mass index (BMI).

| Mild | BMI >16.99 kg/m sq |
| Moderate | BMI 1616.99 kg/m sq |
| Severe | BMI 1515.99 kg/m sq |
| Extreme | BMI < 15 kg/m sq |

### Table 2: DSM5 Diagnostic Criteria for Bulimia Nervosa

A. Recurrent episodes of binge eating. An episode of binge eating is characterized by both of the following:
   1. Eating, in a discrete period of time (e.g., within any 2hr period), an amount of food that is definitely larger than what most individuals would eat in a similar period of time under similar circumstances.
   2. A sense of lack of control over eating during the episode (e.g., a feeling that one cannot stop eating or control what or how much one is eating).

B. Recurrent inappropriate compensatory behavior in order to prevent weight gain, such as self-induced vomiting; misuse of laxative, diuretics, or other medications; fasting; or excessive exercise.

C. The binge eating and inappropriate compensatory behaviors both occur, on average at least once a week for 3 mos.

D. Self-evaluation is unduly influenced by body shape and weight.

E. The disturbance does not occur exclusively during episodes of anorexia nervosa.

Specify if:
- In partial remission: After full criteria for bulimia nervosa were previously met, some, but not all, of the criteria have been met for a sustained period of time.
- In full remission: After full criteria for bulimia nervosa were previously met, none of the criteria have been met for a sustained period of time.

Specify current severity:
- **Mild:** An average of 13 episodes of inappropriate compensatory behaviors per week.
- **Moderate:** An average of 47 episodes of inappropriate compensatory behaviors per week.
- **Severe:** An average of 813 episodes of inappropriate compensatory behaviors per week.
- **Extreme:** An average of 14 or more episodes of inappropriate compensatory behaviors per week.

### Table 3: DSM5 Diagnostic Criteria for Avoidant/Restrictive Food Intake Disorder

A. An eating or feeding disturbance (e.g., apparent lack of interest in eating food; avoidance based on the sensory characteristics of food; concern about aversive consequences of eating) as manifested by persistent failure to meet appropriate nutritional and/or energy needs associated with one (or more) of the following:
   1. Significant weight loss (or failure to achieve expected weight gain of faltering growth in children).
   2. Significant nutritional deficiency.
   3. Dependence on enteral feeding or oral nutritional supplements.
   4. Marked interference with psychosocial functioning.

B. The disturbance is not better explained by lack of available food or by an associated culturally sanctioned practice.

C. The eating disturbance does not occur exclusively during the course of anorexia nervosa or bulimia nervosa, and there is no evidence of a disturbance in the way in which one's body weight or shape is experienced.

D. The eating disturbance is not attributable to a concurrent medical condition or not better explained by another mental disorder. When the eating disturbance occurs in the context of another condition or disorder, the severity of the eating disturbance exceeds that routinely associated with the condition or disorder and warrants additional clinical attention.

7. Other eating disorders
8. Eating disorder, unspecified

**Great Ormond Street classification**
- 1. Food Avoidant Emotional Disorder (FAED)
- 2. Selective Eating
- 3. Functional Dysphagia
- 4. Anorexia Nervosa
- 5. Bulimia Nervosa

**Screening of eating disorders**
Pediatricians can screen for eating disorders
at their office practice, as well as during school health checkups or during sports or similar activities. During health checkups in the Well baby clinics Height, Weight and BMI should be recorded and plotted in the growth charts regularly. If there is any deviation or falloff, it should be treated with suspicion.

SCOFF questionnaire is very simple and short screening tool for EDs, which is used widely by investigators. Though designed originally for adults, it can also help detecting EDs in children and adolescents.

The SCOFF questionnaire is a 5 item questionnaire developed by Morgan et al. The acronym derives from the initials of the five main words/concepts of each of its questions:

(a) Do you make yourself Sick because you feel uncomfortably full?
(b) Do you worry you have lost Control over how much you eat?
(c) Have you recently lost more than one stone (6.35 Kg) in a 3 month period?
(d) Do you believe yourself to be Fat even when others say you are too thin?
(e) Would you say that Food dominates your life?

But studies have found that SCOFF questionnaire may be low sensitivity with high false negatives though they can be highly specific.

ChEat is a modified version of Eating Attitude Test (EAT) for children developed by Garner and Garfinkle. It contains 26 items and another useful screening tool for EDs in children.

Eating Disorder Examination (EDE), is another screening tool, which may be more reliable, as it is an investigator based interview. The latest version of EDE is available, with changes made according to DSM5.

Recently, The Eating Disorders in YouthQuestionnaire (EDYQ), a 14 item instrument for assessing earlyonset restrictive eating disturbances in 8-13 year old children via Selfreport has been formulated by van Dyke et al [14]. The items are based on the criteria for ARFID in the DSM5. EDYQ is reported to be having good psychometric properties, including adequate discriminant and convergent validity.

Sometimes many adolescent girls express concerns about being overweight and may confess about dieting. However most of these children and adolescents may not have an eating disorder. Paradoxically, most patients with eating disorders often try to hide their illness. Hence denials by the adolescent do not exclude the possibility of an eating disorder. Involving parents in history taking may help to identify abnormal eating attitudes or behaviors.

DeSocio et al have developed ‘screening questions for parents’ to be asked by pediatricians to the parents for screening EDs as given below.

1. Have your child’s eating habits changed recently? Yes/ No
2. Do you have any concerns about your child’s weight? Yes/ No
3. Has your child expressed any concern about his weight? Yes /No
4. Has the amount of time your child spend exercising has changed Yes/ No
5. Does your family eat dinner together at night at least 4 times a week?
6. What did your child eat for dinner last night?

If the answer is “yes” to any of the question, then question no 5 and 6 are
asked and further screening for eating disorders is done.

Concerns of parents regarding eating pattern of their children is ubiquitous in civilized world. All Pediatricians see children for feeding difficulties quite commonly. However a good number of them may have misperceived feeding problems. Those with real problems, majority would of them would have milder feeding difficulties. Only few of them can be grouped under ARFIDs. Feeding difficulties (Table 4) can again be classified into Limited Appetite, Selective Intake, Fear of Feeding and problem with the feeding style (Responsive, Controlling, Indulgent or neglecting)\(^{19}\). A tool known as the Identification and Management of Feeding Difficulties for Children (IMFeD) has also been developed and found to be effective in Indian Children in a recent study\(^{20}\).

<table>
<thead>
<tr>
<th>Table 4: Classification of Feeding Difficulties(^{19,20})</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Limited Appetite: These children have actual/misperceived generalized limited appetite. There can be 4 subtypes</td>
</tr>
<tr>
<td>1. Normal child with Misperceived Limited Appetite</td>
</tr>
<tr>
<td>2. Vigorous child with little interest in feeding</td>
</tr>
<tr>
<td>3. Depressed child with little interest in eating</td>
</tr>
<tr>
<td>4. Poor appetite due to organic diseases</td>
</tr>
<tr>
<td>B. Selective Intake: These children consistently avoid specific food of particular taste, texture, smell or appearance.</td>
</tr>
<tr>
<td>1. Misperceived (Neophobia)</td>
</tr>
<tr>
<td>2. Mildly selective</td>
</tr>
<tr>
<td>3. Highly selective (Autism)</td>
</tr>
<tr>
<td>4. Organic cause</td>
</tr>
<tr>
<td>C. Fear of Feeding: These children have intense resistance to feeding, cry or refuse to open mouth at the sight of food.</td>
</tr>
<tr>
<td>1. May be due to Misperceived pain, such as colic</td>
</tr>
<tr>
<td>2. Noxious feeding experience, such as choking or forced feeding</td>
</tr>
<tr>
<td>3. Organic</td>
</tr>
<tr>
<td>D. Inappropriate Feeding Styles: These children may be affected by the parental feeding styles. There are 4 well described feeding/parenting styles, which can affect children’s feeding in positive or negative way. The 1st style of Responsive feeding should be preferred to other 3, which have negative consequences.</td>
</tr>
<tr>
<td>1. Responsive</td>
</tr>
<tr>
<td>2. Controlling</td>
</tr>
<tr>
<td>3. Indulgent</td>
</tr>
<tr>
<td>4. Neglectful</td>
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</tbody>
</table>

Further reading:

1. Srimad Bhagavatam; Book 11, Chapter 8, Text 21.


IAP GDBP Chapter is privileged to be associated with the National Workshop Development for All: Cradle to Crayons and Beyond
Under IAP Action Plan 2016 initiated by Dr Pramod Jog, President

The module contains:

- Developmentally supportive care in NICU
- Red flag signs and development assessment
- International guide for monitoring child development
- ADHD
- Early stimulation workstations
- Emotional Quotient
- Neuromotor impairment

Normal development
High risk neonate
Learning disability
Autism
Behavioral problems
Setting up of an child development centre
Office Approach: Evaluation of Short Stature and Growth Failure

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Introduction
Short stature is probably the most common complaint that brings a child to a pediatric endocrinologist. The majority of patients referred with short stature have a variation of normal growth rather than organic disease. Growth is a general indicator of a child’s health and can be considered a bioassay of the state of health of the child. Growth failure rather than indicating an endocrine abnormality, may herald the onset of non-endocrine systemic disease or a state of malnutrition. Although retarded linear growth rate and delayed pubertal development in a well nourished child may strongly indicate an underlying endocrine basis such as growth hormone deficiency, hypothyroidism, hypercortisolism, and various genetic syndromes. It is imperative that a systematic clinical evaluation coupled with appropriate screening tests as outlined below be undertaken to arrive at an accurate diagnosis and appropriate remedial measures be instituted to prevent any lasting deficits.

Definitions

**Short stature:**
Height less than 3rd percentile (– 2 standard deviation score)

**Growth failure:**
Crossing of 2 percentile lines between the age of 2 years and puberty

Growth velocity less than -1 standard deviation score. (< 25%)

**Childhood growth disorders**

1. **Endocrine causes**;
   (a) Growth hormone deficiency (GHD)
   (b) Hypothyroidism
   (c) Cushing’s syndrome
   (d) Parathyroid or vitamin D disorders
   (e) Hypogonadism

2. **Non-endocrine causes**:
   (a) Syndromes (Turner, Noonan, Russell-Silver, etc)
   (b) Chronic disease; malnutrition, inflammation, psychosocial short stature
   (c) Intrauterine growth retardation (IUGR, SGA)
   (d) Chronic glucocorticoid Usage
   (e) Familial (genetic) short stature (FSS)
   (f) Constitutional delay of growth and puberty (CDGP)

3. **Idiopathic short stature (ISS)**

**Clinical approach to short stature and growth failure**

**History:**
(a) Age of onset of growth failure
(b) School, home or physician records of previous heights and weights must be sought and charted on BMI charts, distance and velocity growth charts. Birth weight, gestational age, Family history of constitutional short stature. Developmental milestones

(b) Behavioral history.

(c) Nutritional history

(d) Medication History: prolonged intake of steroids, dextroamphetamines.

(e) Edema hands and feet at birth is suggestive of Turner's syndrome

(f) Breech delivery, neonatal hypoglycemia, jaundice, micropenis may indicate multiple pituitary hormone deficiencies

(g) Family history: short stature, delayed puberty in parents may indicate familial short stature, constitutional delay in growth

(h) Short stature or a reduced rate of linear growth in a child with obesity suggests the possibility of growth hormone deficiency, hypothyroidism, cortisol excess, pseudohypoparathyroidism, or a genetic syndrome such as Prader-Willi syndrome.

(i) Dry skin, constipation, intolerance to cold, or fatigability suggests hypothyroidism.

(j) Chronic shortness of breath may suggest acyanotic congenital heart disease, asthma bronchiectasis or cystic fibrosis.

(k) Fever, night sweats, anorexia, weight loss may indicate tuberculosis.

(l) Chronic diarrhea, steatorrhea and abdominal pain suggest malabsorption

(m) Polyuria and polydipsia may be noted in renal tubular acidosis, diabetes insipidus.

(n) A history of damage to the CNS (infection, trauma, hemorrhage, radiation therapy, seizures) with growth failure suggests pituitary growth hormone deficiency or pituitary hypothyroidism.

(o) A history of morning headaches, vomiting, visual disturbances, and excessive urination or drinking also suggests that the growth failure may be caused by a tumor or mass in the hypothalamus.

**Examination**

Anthropometric measurements: weight, standing height, upper to lower segment ratio, must be taken. Weight height, BMI should be charted and growth velocity should be recorded. Pubertal development to be assessed by Tanner's sexual maturity rating. Stretch penile length and testicular size to be recorded. Measurement of height should be done on a well calibrated stadiometer / infantometer. Height velocity is measured over a period of 6 to 12 months.

**Normal growth velocity:**

In the first year of life a child grows by 25cm, 12.5 cm in 2nd year, 6-7cm in 3rd and 4th year, 5cm per year from 5-9 years with a nadir of 3.5 cm per year in prepubertal age group. During pubertal growth spurt 10-30 cm height is gained, with peak height velocity of 9-11 cm per year in boys and 7-9 cm per year in girls.

Upper to lower segment ratio helps to differentiate between proportionate and disproportionate causes of short stature.

Body proportions (upper segment: lower segment) change from 1.7 at birth to 0.98-1 by 13-14 years of age and to 1 in adulthood.

Plot the height against mid parental height range. Mid parental height (MPH) is calculated by adding 6.5cm to the average of mothers and fathers height in boys and by subtracting...
6.5 cm in case of girls. This should be plotted on growth chart with a range of about 8.5 cm below or above MPH. If a child lies within this range he has a genetic cause of short stature. Calculation of weight for height is helpful in differentiating wasting (malnutrition, systemic illnesses), obesity (Cushing’s syndrome) and stunting (GHD). Two particularly useful clinical measures are the rate of linear growth and the timing of puberty. Growth rate and pubertal development are diminished or delayed in growth hormone deficiency, hypothyroidism, cortisol excess, and various genetic syndromes.

Thorough general and systemic examination needs to be performed to look for dysmorphism (congenital syndromes) goiter, dry skin (hypothyroidism) acanthosis nigricans, purple striae, hypertension, central obesity (Cushing’s Syndrome) cherubic facies, frontal bossing, malar hypoplasia, depressed bridge of nose (growth hormone deficiency) Frontal bossing, beading, wrist widening (rickets) Hypertension (chronic renal failure) Round face, short 4th metacarpal, mental sub normality (pseudohypoparathyroidism) Disproportionate short stature (skeletal dysplasia, rickets, hypothyroidism) Vitamin deficiency signs (PEM malabsorption) Midline defects central incisor (hypopituitarism)

**Investigations**

**Bone age:**

Bone age is done to study the skeletal maturity. It is a reflection of the physiologic age rather than the chronologic age of the child. IT IS DONE BY TAKING HAND AND WRIST X-RAY OF LEFT HAND

Two systems for reading bone ages are available-Greulich and Pyle’s Atlas method (American) and Tanner and Whitehouse scoring method (European). With a bone age over 6 years and an accurate determination of stature, final height can be predicted by the Bailey-Pinneau tables found in the Greulich and Pyle Atlas. The Roche Wainer Thissen (RWT) method allows prediction of eventual adult height in younger children.

**Familial short stature:**

H.A< B.A=C.A; Constitutional growth delay: H.A=B.A<C.A

If height is normal for national standards and mid-parental height and growth velocity is normal on, follow up then reassurance is needed without any further investigations. Normal bone age at outset usually rules out pathological cause of short stature.

**Laboratory studies**

**Screening tests:**

CBC, Hb, ESR (anemia, infection), X-ray Left wrist and Hand (Bone age, Rickets) Free T4 and TSH (Hypothyroidism) Renal, Liver function test (CRF, CLD) Total proteins, albumin (Nutrition) Serum Ca, P, alkaline phosphatase (rickets, pseudohypoparathyroidism) Blood gas, serum electrolytes ( metabolic acidosis, RTA) Urine routine microscopy, pH (RTA, chronic pyelonephritis, glomerulonephritis) Stool routine microscopy (malabsorption, giardiasis) Celiac screen, malabsorption workup IGF1, IGFBP3 (GHD) Karyotype (Turner, Trisomy)
**Diagnostic approach to short stature (Important messages)**

1. Careful height measurements and puberty assessment are very important.
2. Variations of normal are much commoner than true pathological conditions.
3. The children who require investigation are the ones who are growing abnormally slowly (Subnormal growth velocity).
4. Approach the problem of short stature as general paediatricians and diagnose occult systemic illness.
5. True endocrine causes are rare. Subnormal growth velocity, delayed bone age in overweight children are suggestive of endocrine causes.
6. Early diagnosis of pathological conditions is imperative.
7. Remember Turner syndrome and analyse the chromosomes in short females.

Evaluation of short stature starts with serial measurements of linear growth. Growth faltering should be addressed by growth specialists and should be detected early by the pediatric specialist.

(BA- Bone age, CA – Chronological age, HA- Height age, CDGP- Constitutional Delay in Growth and Puberty, RTA- Renal Tubular Acidosis, CRF- Chronic renal failure, CLD- Chronic Liver Disease GHD- Growth Hormone Deficiency).
Case Report - Cockayne Syndrome

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Abstract
Cockayne syndrome is a rare neurodegenerative disorder with an autosomal recessive inheritance. Patients with this syndrome present with global developmental delay, growth failure, photosensitivity, mental retardation and dysmorphic features. This is due to two gene mutation of ERCC 8 and ERCC 6 located on chromosome 5 and 10.

In this case, a six-year-old male child presented with global developmental delay, spastic diplegia, growth retardation and dysmorphic features. CT scan of brain showed bilateral basal ganglia calcification. NCV study showed peripheral neuropathy. Child had bilateral hearing loss. Ophthalmological examination revealed salt and pepper retinopathy and hypermetropia. Gradually there was development of unique gait due to contracture in both lower limbs. Associated dental decay was found in the child and provisional diagnosis of Cockayne syndrome was made. Gene mutation study was advised.

It’s a rare disorder, incidence is 2 per billion babies worldwide and it has a poor prognosis.

Key words: Cockayne syndrome, global developmental delay, premature ageing

Introduction
Cockayne syndrome is a rare neurodegenerative disorder with an autosomal recessive inheritance. Patients with this syndrome present with impaired development of the nervous system, global developmental delay, growth failure, photosensitivity, eye disorders, dysmorphic features and premature ageing. It is also known as Neill-Dingwall syndrome. Failure to thrive and neurological disorders are criteria for diagnosis, while photosensitivity, hearing loss, eye abnormalities are other very common features. Problems with any or all of the internal organs are possible.

It is associated with a group of disorders called leukodystrophies, characterized by degradation of white matter. The underlying disorder is a defect in a DNA repair mechanism. It is named after English physician Edward Alfred Cockayne who first described it in 1936.

Case report
Six-year-old male child came with clinical features of global developmental delay including delay in speech and hypertonic limbs. He was second child born out of non-consanguineous marriage, born at term by normal vaginal delivery with birth weight of 3 kg. There was no history of birth asphyxia. There was no history of neonatal seizure or hyperbilirubinemia. The child had achieved
head control at 1 year, sitting independently at 2 years, standing with support at 2 ½ years, standing without support at 3 years and bisyllable words at 3 years (Fig 1 & 2). Dysmorphic features like microcephaly, prominent cupped ear, deep set eyes, synophrys, crowded and carries teeth with thick lips were present. Growth was stunted with thinning of skin and hair. Anthropometric measurements like OFC, height, weight were below 3rd centile for age.

**Developmental assessment:**
He could sit and stand without support, manipulate objects, scribble with his right hand and able to speak bisyllables words. He could follow simple requests and was orientated to his surroundings. He showed good interest in music based activities. Indicated his needs through facial gestures with vocalization.

Motor skill corresponds to 1 year 2 month, socialization skill corresponding to about 10 months and Language skills 11 months.

**CNS examination:**
Child was alert, friendly, with bi-syllable speech and no meaningful words. He was right handed and cranial nerves were intact.

*Motor system* – Bilateral contractures of achilles tendon, increased tone with symmetric atrophy. Power grade 4/5 (right side) and 5/5 (left side). Involuntary movements were absent.

Reflexes – Superficial reflexes were absent and deep tendon reflexes were brisk.

**Investigation**

**CT scan brain:**
Bilateral calcification of putamen and subtle calcification of frontal parietal cortex at depth of sulci and bilateral calcification in dentate nuclei (Fig 3).
**MRI brain:**
Bilateral chunky calcification in basal ganglia with diffuse calcification in frontal and parietal regions. Diffuse cerebellar and subcortical volume loss in brainstem, with diffuse white matter volume loss including the corpus callosum (Fig 4 & 5).

**MR spectroscopy:**
It was done to detect neurometabolites. It showed decreased NAA/creatinine and choline/creatinine ratio suggestive of Cockayne syndrome.

**Sleep EEG:**
Normal

**EMG:**
Sensory motor de-myelinating and axonal polyneuropathy in all limbs.

**Evoked potential report (SSEP report):**
Bilateral dorsal cord dysfunction.

**Ophthalmological examination:**
Salt and pepper retinopathy with hypermetropia.

**Brainstem auditory evoked response and DPOAE:**
No response bilaterally (central pathology).

**Diagnosis**
This child presenting with the clinical and investigative features of typical dysmorphism, developmental delay, hearing loss and eye findings, classical neuroimaging findings, point towards Cockayne syndrome with sensory, motor demyelinating and axonal polyneuropathy.

This is diagnosed as a case of CS TYPE 1 as he satisfies 2 major criterions and 3 minor criterions, besides age of onset.

**Major criterion** – Abnormal postnatal growth, progressive microcephaly and neurologic dysfunction followed by behavioral and intellectual decline

Minor criterion - Hearing loss, dental decay and cachectic dwarfism, photosensitivity.

For diagnosis in a small child 2 major criterion is sufficient for diagnosis. In older children, 2 major criterion with 3 minor criteria are required for diagnosis.

Gene mutation study was advised to facilitate genetic counselling.

**Discussion**
Cockayne syndrome (CS) is a rare condition with about 40 cases reported in world literature. The underlying disorder is a defect in a DNA repair mechanism. Unlike other defects of DNA repair, patients with CS are not predisposed to cancer or infection. Cockayne syndrome is a rare but destructive disease usually resulting in death within the first or second decade of life.

The mutation of specific genes in CS is known. This is due to two gene mutation of ERCC 8 and ERCC 6 located on chromosome 5 and 6.
10. But the widespread effects and its relationship with DNA repair is yet to be well understood.

There are three recognized categories of CS, depending on age of onset, type 1, 2 and 3.

**CS type I:**
The "classic" form, is characterized by normal fetal growth with the onset of abnormalities in the first two years of life. Vision and hearing gradually decline. The central and peripheral nervous systems progressively degenerate until death in the first or second decade of life as a result of serious neurological degradation. Cortical atrophy is less severe in CS Type I.

**CS type II:**
It is present from birth (congenital) and is much more severe than CS Type I. It involves very little neurological development after birth. Death usually occurs by age seven. This specific type has also been designated as cerebro-oculo-facio-skeletal (COFS) syndrome. Typically, patients with this early-onset form of the disorder show more severe brain damage, including reduced myelination of white matter, and more widespread calcifications, including in the cortex and basal ganglia.

**CS type III:**
It is characterized by late onset, is typically milder than types I and II. Often patients with type III will live into adulthood.

Persons with this syndrome have microcephaly, dwarfism, sunken eyes and they have an "aged" look. They often have long limbs with joint contractures, kyphosis, and they may be cachectic, due to a loss of subcutaneous fat. Their small chin, large ears, and pointy, thin nose often give an aged appearance. The skin of those with Cockayne syndrome is also frequently affected. Hyperpigmentation, varicose or serious sensitivity to sunlight is common. Often patients with Cockayne syndrome will severely burn or blister with very little exposure. The eyes of patients can be affected in various ways and eye abnormalities are common in CS. Cataracts and corneal opacity is common. Damage to optic nerve, causing optic atrophy can occur. Nystagmus and pupils that fail to dilate demonstrate a loss of control of voluntary and involuntary muscle movement. A salt and pepper retinal pigmentation is also a visible symptom.

Diagnosis is determined by a specific test for DNA repair, which measures the recovery of RNA after exposure to UV radiation.

**Prognosis**
CS TYPE 1 typically dies within 1st or 2nd decade of life. All reported cases have cachectic dwarfism. Neuropathological changes include neuronal and myelin loss with deposits of calcium and iron in the vessels of cerebellum, basal ganglia and cerebrum.

**Treatment**
There is no permanent cure for this syndrome, although children can be treated according to their specific symptoms. The prognosis for those with Cockayne syndrome is poor, as death typically occurs by the second or third decade of life.

Treatment usually involves physical therapy and minor surgeries to the affected organs, like cataract removal. Also wearing high-factor sunscreen and protective clothing is recommended as patients with Cockayne syndrome are very sensitive to UV radiation. Optimal nutrition can also help.

Genetic counselling for the family is
recommended, as the disorder has a 25% chance of being passed to any future children. Another important aspect is prevention of recurrence of CS in other sibling. Identification of gene defects involved makes it possible to offer genetic counselling and antenatal diagnostic testing to parents who already have one affected child.

Further reading:

Please be a Member of IAP GDBP

All members of Indian Academy of Pediatrics can be a life member of IAP Chapter on Growth, Development and Behavioral Pediatrics. We request the members of IAP in various States to kindly be a member of this chapter by filling the following form and deposit it in the address of the secretary with a DD of Rs.550/- in favour of “Growth, Development and Behavioural Pediatrics Academy” payable at Kolkata. Membership Form Send it to The Secretary, GDBP Chapter “Oriental Apartments” Flat H1, 15C, Canal Street, Kolkata 700 014.

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5. Corresponding Address : ........................................................................................................................................

6. Designation in Institute with its Name (if any) ..............................................................

7. Academic Qualification (s) :.................................................................................................

8. Phone No. Residence :..............................Office/Chamber :................................................
9. Mobile No. :............................................10. Email :.................................................................

11. Bank Draft/Cheque No:..............................Name of Bank:..............................
    Branch:.................................................................................................................................
    Date                                                                                   Signature

IJGDBP
Ethical Issues in Pediatrics

Jaydeep Choudhury
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Ethics is a quest for wisdom. In literally term ethics refers to the study or the science of morals, the system or set of principles particular to a certain person, community or group. Bioethics is the process of determination of the most morally desirable course of action in health care given the conflicting values inherent in varying treatment options of a particular condition.

Pediatrician has an independent professional obligation to act in a child’s “best interest”. But at the same time pediatrician is at the juncture of the child and the parents. The scope of parental decision making is governed by the child’s best interest and the emerging capacity of autonomy of an older child or adolescent. Thus there is a possibility of conflict between the pediatrician, the child and the parents. Arguably the role of the pediatrician is pivotal. The pediatrician under the circumstances is the competent person and is the best judge of the treatment needed. He has the dual responsibility of judging the condition and explaining the treatment options and possible outcome to the receiver. Each situation has to be approached with respect to the parent’s responsibility for the wellbeing of the child and the child’s developing capacity and autonomy. The prevailing social milieu, cultural acceptability and option of alternative approaches to health care add to the complexity of decision making.

A pediatrician enters a professional relationship with a child in two ways, either in elective or emergency situation. In the former the dealing is relatively simpler. One gets the time and opportunity to interact and explain. The parents are usually more receptive in these situations also. In an emergency setting the complexity arises. Here the parents are usually confused. A pediatrician often has to act by instinct. Often a life saving procedure like tracheostomy has to be undertaken with minimal explanation. Otherwise the child’s life may be at risk.

A pediatrician has the responsibility to provide the following medical care:

1. Emergency care to all patients coming to an emergency set up
2. Ensure that competent medical care is provided
3. Give proper advice to the “best interest” of the child

The relationship thus entered may be terminated in the following circumstances:

1. When the patient has recovered and does not require care
2. Parent or guardian and in cases of older child or adolescent himself or herself agrees to terminate the relationship
3. A proper transfer of the patient has been arranged.
The most recent attempt to develop a widely (internationally) applicable bioethics code is the adoption of the Universal Declaration of Bioethics and Human Rights in October 2005 by the United Nations Educational, Scientific, and Cultural Organization.

The general moral obligations in medical decision making include the following:

(a) Autonomy – Respect the wishes of the person who is competent
(b) Nonmaleficence – Do not harm others
(c) Beneficence – Benefit others
(d) Usefulness – Produce a net balance of benefit over harm in all the actions
(e) Justice – Distribute benefits and harm of a particular condition fairly
(f) Fidelity – Keep promises and contracts
(g) Veracity – Truthfulness and disclosure should be adhered to
(h) Confidentiality – Protection of private information is of utmost importance.

Role of parents in ethical decisions

In pediatrics, a new level of complexity is added. Decision that may affect the subject’s future employability, insurability, privacy and other interests are made by surrogates (usually parents).

Under all circumstances, the decision which is considered best for the child must be balanced against considerations which are best for the family. Pediatricians should respect the wish, socio-cultural interests and the resources of the parents. The interest of the sibling has to be considered also. An all-out effort to save a child if turns futile and the parents exhaust all their resources may adversely affect the future life of the sibling. In such a situation the parents have to be explained and their wish should be considered appropriately. Parental divorce is often high in such situations. These circumstances may precipitate sibling maladjustment, educational lag, delinquency and behavioural problems.

Presence of family during procedures:

The question whether the family should be present during invasive procedures and especially cardio-pulmonary resuscitation is debatable. The whole may be simplified to some extent if one health worker is provided to counsel them.

Emergency care:

When a child is brought to the emergency setup the consent for treatment is implied. Even if the parents are not present the child should be provided appropriate life-saving treatment.

Religious considerations:

Sometimes religious interests may hinder the decision of best treatment of the child as perceived by the pediatrician. A particular community refuse to accept blood transfusion under any circumstances. They ask for all other medically and scientifically based alternatives. The ethical responsibility of the treating paediatrician is tested under these situations. Forcible imposition of a treatment modality often fails. It is best to persuade, explain and trust the family.

Divorced parents:

The custodian parent is legally competent to give consent for the child’s treatment.

Disclosure of information to children:

Age-appropriate information should be provided to the children in an appropriate manner in order to help them to participate in decision making. One such situation is circumcision, which is sometimes done on religious and socio-cultural ground. It may be considered unethical.
as a part of the skin of the child is taken away without consent and the procedure has doubtful benefit. Malignancy and HIV infection in a child is often a critical issue. Parents hesitate to share information with the child. Pediatrician should judge accordingly and understand parental feelings. The parents can be reassured that the child can cope up better when truthfulness and trust are maintained.

Conflicts of ethical principles
Pediatrician is faced with a tricky situation when there is a conflict between the following:
1. Parents’ rights and the child’s rights
2. Child’s rights with pediatrician’s duties
3. The interest and responsibility of parents and paediatrician versus that of community.

Institutional Ethics Committee (IEC)²
An IEC usually performs the following three different functions:
(i) Drafting and review of institutional policy on the ethical issues like life-sustaining medical treatment (LSMT) or do not attempt resuscitation (DNAR).
(ii) Education of health care professionals, parents and families about ethical issues in health care.
(iii) Case consultation.
The basic aim of the art and science of medicine is to help people escape the evils of disease, handicap and death. At the same time it is the duty of the physician to help with newer advances and technology. An ideal doctor is one who goes one step ahead and stands beside the patient in escaping the evil even when the disease is incurable, the handicap inevitable and unbearable and death is the culmination. At times a “good” remedy from medical standpoint is worse than the disease. The doctor should be flexible. The ideal medical treatment in certain situations has to be weighed against the ethical considerations. After all being a physician and more so a pediatrician is a complex, critical and often demanding job.

Further reading: