CEREBRAL PALSY PRESENTING AS RECURRENT PNEUMONIA

Mubarak Ali[1], Abida Mubarak[1], MI Babar[1], Shahzad Ali Bukhari[1], Javaria Yunas[1]

ABSTRACT

Background: Cerebral palsy is a common cause of childhood morbidity and mortality particularly in developing countries like Pakistan. Though it usually presents as gross motor developmental delay, however, one of its common but under reported mode of presentation is recurrent pneumonia. **Objective:** To determine the clinical and radiological profile of children with cerebral palsy presenting as recurrent pneumonia. **Patient & Methods:** This prospective descriptive study was conducted at a tertiary care centre for a period of one year, from 1st January to 31st December 2011. All the patients who presented with recurrent chest infections were evaluated for their neurodevelopmental status with the help of detailed history and meticulous examination. Only those cases of recurrent pneumonia who were also having cerebral palsy were included in the study while all neurodevelopmentally normal or those having neurodevelopmental abnormalities other than cerebral palsy were excluded from the study. In all cases of suspected cerebral palsy detailed history especially regarding developmental milestones, complaints of respiratory system (fever, cough, dyspnea, tachypnea etc.), birth history, feeding history and family history was taken. The symptoms of overt direct aspiration i.e. coughing, chocking, gagging, apnea, becoming dyspnoeic / tachypnoeic / cyanotic during feeding or restlessness after feeding were specifically asked and feeding was also observed. Meticulous clinical examination including detailed examination of the oral cavity, respiratory, cardiovascular and central nervous system was carried out. Developmental and CNS examination was also repeated after recovery from pneumonia. In infants extensor spasm of neck & trunk muscles, demonstrated by both ventral and dorsal suspension was the most sensitive sign for the diagnosis of cerebral palsy. Investigations carried out in all patients were CBC and X-Ray chest while blood C/S, serum biochemistry, Arterial Blood Gases (ABGs), ECG, echocardiography and CT scan brain was carried out in selective cases where indicated. **Results:** During the study period of one year, 14805 patients were admitted in Pediatric department. Of these, 4442 (30%) were neonatal and 10363 (70%) were post-neonatal admissions. Recurrent pneumonia as a result of cerebral palsy was diagnosed in 168 patients who accounted for 1.13% and 1.62% of the total and post-neonatal admissions respectively. 108(64%) patients were below the age of one year, 45 (27%) patients from 1 to 5 years while 15 (09%) patients were having age above 5 years. Radiological findings were also variable among different age groups with involvement of both lungs in almost all the cases, main findings included the presence of generalized hyperinflation along with wide spread, particularly perihilar infiltrates and consolidation / collapse, mainly of right upper / middle lobes. **Conclusion:** Cerebral palsy is a common disorder leading to disability and deaths. Respiratory problems especially recurrent pneumonia is a common complication of cerebral palsy. Solid and sustainable strategies need to be developed and implemented to reduce the incidence and complications of cerebral palsy.

Key Words: Cerebral palsy, aspiration pneumonia, recurrent pneumonia, gastroesophageal reflux (GER).

INTRODUCTION

The term cerebral palsy (CP) was initially coined more than a century ago considering the condition as “brain paralysis." More precisely cerebral palsy is defined as a group of non-hereditary disorders of the movement and posture resulting from non-progressive brain lesions incurred during the period of developing fetal or infant brain i.e; up to the age of 3 years. However, it is not a single diagnosis but an "umbrella" term collectively describing non-progressive brain lesions involving motor or postural abnormalities noted during early development.[1] Nevertheless, regardless of the etiology and the changing clinical picture overtime, the underlying brain lesion in cerebral palsy is static; hence, cases associated with underlying disorders of progressive or degenerative nature are excluded when diagnosing cerebral palsy.[2]

Though the usual mode of presentation of cerebral palsy is gross motor developmental delay, however, it may present with a wide variety of clinical manifestation or complications/associations affecting multiple systems, more particularly respiratory system.[3] The etiology of respiratory complications in children with cerebral palsy is multi-factorial; in fact, several of these factors co-exist and may interact with each other to compromise the quality of life in these already handicapped children. These factors include recurrent aspiration, poorly functioning mucocilliary accelerator, ineffective cough reflex, weak musculature, lack of exercise / physical activity, malnutrition and recurrent infections other than respiratory system to which these individuals are also more prone. Children with cerebral palsy often appear to have ineffective cough reflex due to weak musculature

1. Paediatrics Department, Sheikh Zayed Medical College/Hospital, Rahim Yar Khan.

Correspondence: Prof. Dr. Mubarak Ali, Head Department of Paedaratrics, Sheikh Zayed Medical College/Hospital, Rahim Yar Khan.

Email: mubarakalidr@hotmail.com
Mobile: 0300-8776655

and diminished sensitivity to cough, perhaps as a result of desensitization of airway irritant receptors because of chronic/recurrent aspirations resulting in "silent" aspiration.[4,5] In healthy children, exercise induces deep breathing which aids clearance of secretions and opens up under ventilated lung regions, the child with cerebral palsy unable to take part in vigorous exercise, is prone to atelectasis and subsequent superimposed infection.[6]

Aspiration, the accidental entry of a foreign substance (solid & liquid even saliva) into the respiratory tract, occur frequently in many healthy individuals in very small quantities but the normal defense mechanisms particularly cough reflex and mucocilliary accelerator remove the aspirated material without any adverse effects. In cerebral palsy not only these normally active "sweep up" local defense mechanisms become sluggish but also there is recurrent aspiration due to muscle weakness, dystonia, poor coordination and high incidence of gastroesophageous reflux.[7] Recurrent aspiration in cerebral palsy is both direct & indirect. Direct aspiration occurs directly from the oral cavity including feed / food materials (liquids & solids) and oral and upper respiratory secretions into lower airways due to neuromuscular in-coordination and inadequate protective reflexes. Failure of proper bolus formation, oesophageal peristalsis, glottis closure, and "turn taking" between swallowing and breathing leads to recurrent aspiration during feeding. Thin liquids are particularly prone to be aspirated. Even between feeds, there is likely to be recurrent aspiration of non-sterile oral and upper respiratory secretions into the (normally sterile) lower airways because of poor protective reflexes[8]. In indirect aspiration, it is the regurgitated stomach contents which are inhaled into the lower airways. For reasons, poorly understood and little researched, GER appears to be more common, persistent, and severe in children with cerebral palsy. This may be partly caused by spasticity of abdominal muscles causing increased intra-abdominal pressure, but it is likely that in-coordinated oesophageal and sphincter muscle activity also plays a part. The refluxate may not be actively cleared as a result of disturbed oesophageal peristalsis, and is more likely to "travel up" and get aspirated.[9]

In underprivileged countries, malnutrition is a common resultant companion of cerebral palsy. Under feeding, gastroesophageal reflux, recurrent infections and additional energy expenditure all contribute to reach this unfortunate state of affairs. The resulting malnutrition subjects the respiratory muscles to catabolism, leading to atrophy, weakness and reduced lung function, as well as increasing bacterial colonization of the airways and reducing resistance to infection.[10,11] When this dangerous vicious cycle of recurrent infections and malnutrition evolves, it becomes extremely difficult to get rid of it and the usual ultimate outcome is death.

Cerebral palsy remains the leading cause of childhood disability, particularly in developing countries like Pakistan. Infants and young children with cerebral palsy frequently present with complaints pertaining to respiratory system which play a major role in the life quality and expectancy of these children. This common clinical experience though encountered and recognized frequently in day to day practice, is however, not well reported in the literature, especially local literature. The present hospital based study was carried out to determine the clinical and radiological profile of children with cerebral palsy presenting as recurrent pneumonia.

PATIENT AND METHODS

This prospective descriptive study was conducted at the department of Pediatrics, Sheikh Zayed Hospital/ Medical College, Rahim Yar Khan for a period of one year, from 1st January to 31st December 2011. Our hospital is a tertiary care centre with very wide catchment area including District Rahim Yar Khan, District Rajan Pur (across the river Indus) and adjoining districts of the province of Sind and Baluchistan. The following case definitions were applied in the present study.

The demographic data of all children with diagnostic codes corresponding to diagnosis of pneumonia were identified according to the International Classification of Diseases, Ninth Revision; Clinical Modification (codes 480-487, 507). Recurrent pneumonia was defined as 02 episode of radiologically confirmed pneumonia within the same year, or 03 or more episodes over any time period with complete clinical and radiological resolution in between acute episodes.[12] Cerebral palsy was diagnosed clinically by the presence of delay in gross motor milestones, however, in infants extensor spasm of neck & trunk muscles, demonstrated by both ventral and dorsal suspension, was found and

also taken as the most sensitive sign for diagnosis.Aspiration pneumonia was diagnosed clinically by the presence of history of coughing, chocking, gagging, apnea, becoming dyspnoeic / tachypnoeic / cyanotic during feeding or restlessness after feeding. The presence of excessive drooling or pooling of secretions in oral cavity was also considered as a risk factor for aspiration.[5,13] Radiologically, aspiration pneumonia was diagnosed by the involvement of both lungs in almost all the cases, presence of generalized hyperinflation along with wide spread, particularly perihilar infiltrates and consolidation / collapse, mainly of right upper / middle lobes. Moreover, as in some of the cases it was difficult to distinguish consolidation from atelectasis, hence in our study; for the sake of simplicity we have used the word collapse / consolidation for these findings.[14]

In all cases of suspected cerebral palsy detailed history especially regarding developmental milestones, complaints of respiratory system (fever, cough, dyspnoea, tachypnoea and wheezing etc.), birth history, feeding history and family history was taken. The symptoms of overt direct aspiration i.e. coughing, chocking, gagging, apnea, becoming dyspnoeic / tachypnoeic / cyanotic during feeding while restlessness after feeding was specifically asked and feeding was also observed.[7] Clinical features of Gastroesophageal reflux e.g. vomiting, nasal regurgitation were also inquired. Meticulous clinical examination including detailed examination of the oral cavity, respiratory and central nervous system was carried out. Developmental and CNS examination was also repeated after recovery from pneumonia. Investigations carried on all patients were CBC and X-Ray chest while blood C/S, serum biochemistry, ABGs, ECG, echocardiography and CT scan brain was carried out in selective cases.

Inclusion criteria: All the patients who presented with recurrent chest infections were evaluated for their neuro-developmental status with the help of detailed history and meticulous examination. Only those cases of recurrent pneumonia which were also having cerebral palsy were included in the study.

Exclusion criteria: All the patients of recurrent pneumonia who were neurodevelopmentally normal or having neurodevelopmental abnormalities other than cerebral palsy (e.g. Neurodegenerative brain disease, spinal muscular atrophy) were excluded from the study.

RESULTS

During the study period of one year, 14805 patients were admitted in Pediatric unit. Of these, 4442 (30%) were neonatal and 10363 (70%) were post-neonatal admissions. Recurrent pneumonia as a result of cerebral palsy was diagnosed in 168 patients who accounted for 1.13% and 1.62% of the total and post-neonatal admissions respectively. Of these 108 (64%) were males and 60 (36%) were females. 108 (64%) patients were below the age of one year, 45(27%) patients from 1 to 5 years while 15 (09%) patients were having age above 5 years. The youngest case was of 2 months while the age of eldest case was 12 years (Table I). In many of the cases of neurodevelopmental disability, on radiography, there was involvement of more than one lung lobe as shown in the table-II.

Table I: Age groups of patients having recurrent pneumonia along with cerebral palsy (N=168)

Age group	No. of patients	%age
Below 1 year	108	64%
1 - 5 years	45	27%
Above 5 years	15	09 %

Table II: Radiological findings of the patients (N=168)

Characteristics	Age groups		
	Below 1 year	1 – 5 years	Above 5 years
Patients (No)	108	45	15
Generalized hyper inflation	102 (94%)	33 (73%)	06(40%)
Diffuse bilateral mainly perihilar infiltrates	84 (77%)	30 (66%)	09(60%)
Right upper lobe consolidation / collapse	96 (88%)	27(60%)	03 (20%)
Right middle lobe consolidation / collapse	06(5.5%)	15 (33%)	06 (40%)

DISCUSSION

Respiratory tract infections remain the commonest cause of seeking medical advice and pneumonia as the major killer of children especially under the age of 5 years. However, a child with repeated chest infections can pose

a difficult diagnostic challenge for the Pediatrician.[15,16] While many factors contribute to the occurrence of repeated lower respiratory tract infection in children, the role of cerebral palsy, a common cause of repeated pneumonia in our country, in the causation of recurrent pneumonia has not been explored judicially in literature.

Normal mechanisms of safe swallowing include raising of the palate, epiglottic tilt, cord closure, primary peristalsis of the esophagus, and the tone and sequential relaxation of the upper and lower oesophageal sphincters.[13] It is not surprising that in cerebral palsy, this highly and precisely coordinated choreography is profoundly disturbed by muscle weakness, dystonia and poor coordination. Moreover, high incidence of GER and weak local defense mechanisms including sluggish mucocilliary accelerator make these patients prone to have recurrent aspirations.[17] The recurrent aspiration of oral, nasal, or gastric contents can lead to several clinical presentations, including persistent/recurrent bronchitis or bronchiolitis, recurrent pneumonia, atelectasis, bronchiectasis, wheezing, cough and apnea or laryngospasm.[18]

In our study, 168 patients were having recurrent pneumonia as a result of cerebral palsy, of which 108 (64%) were males and 60 (36%) were females. Of these 168 cases, 108(64%) were below the age of one year, 45(27%) from 1 to 5 years while 15 (09%) patients were having age above 5 years.

There were more cases (64%) of recurrent aspiration pneumonia in infancy, as oropharyngeal incoordination in cerebral palsy is more common, severe and persistent in infancy, however it tends to improve with age as does gastroesophageal reflux,[19] hence there was less number of cases after infancy and the number went on decreasing with increasing age. The predominance of younger age group in having recurrent pneumonia also reflects the usual pattern of higher incidence of respiratory infections in younger age group. The infants are 1st time exposed to a variety of new microorganisms with which they have to be exposed in future life. It is pertinent to mention it here that in our study most of cases were either preceded by or were having concomitant intercurrent infections like upper respiratory tract infections, gastroenteritis at the time of presentation. The presence of concomitant infections further compromise the finely coordinated swallowing reflex in these already neuro-developmentally handicapped patients making them more prone to have direct or indirect aspiration. The similar pattern of more cases of aspiration during concomitant upper respiratory tract infections was also observed in other study.[20-23]

The pattern of radiological findings was also quite different in different age groups. The two most common radiological findings in infants were generalized hyperinflation (increased lung volume) and right upper lobe consolidation/collapse present in 102 (94%) and 96 (88%) cases respectively. It was followed by generalized but mainly perihilar infiltrates present in 28 (77%) patients. Right middle lobe consolidation/collapse was present in only 06 cases, similarly; consolidation/collapse of left upper lobe and a few middle segments of left lung were also present in 06 cases each. In infants the most dependant part of the lungs in supine position, which is the usual position during feeding in infants, is upper lobe of right lung. It is followed by middle and lower lobes of right lung and then different lobes of left lung.[18,19]

Among the 45 cases in the age group between 1 to 5 years, generalized hyperinflation was the commonest radiological finding present in 33(73%) patients while 30 (66%) cases had diffuse but mainly perihilar infiltrates. Though 60% cases had right upper lobe and 15 (33%) had right middle lobe collapse /consolidation, the involvement of right upper lobe in this age group was relatively less common as compared to that in infants while radiological findings in right middle lobe was present in more percentage of cases in this age group in comparison to infants. These findings are mainly present in the posterior segments of right upper lobe in infancy (being the most dependent segments in spine position feeding) and posterior segments of right middle and lower lobe after infancy (being the most dependent segments when the baby is fed in upright position).[13]

Among 15 cases in above 05 years age group, 09 (60%) cases had right lower lobe, 06 (40%) cases had right middle lobe and 03 (20%) cases had right upper lobe collapse/consolidation, however, amongst these, in one case both right middle and lower lobes were involved while in another 2 cases lower lobes of both lungs were involved . Nine (60%) cases had diffuse but mainly perihilar infiltrates while generalized hyperinflation was present in 06 (40%) cases. Though diffuse infiltrates were

frequently seen but lung volume was not markedly increased in comparison to the younger age groups.

In a series of 238 children hospitalized with recurrent pneumonia, the most common underlying problem was oropharyngeal incoordination present in 48% of the patients.[18] In another study 34% of children with neurodevelopmental delay had recurrent pneumonia.[8] 24 out of 34 have recurrent aspiration pneumonias in another study.[21] Abdullah F et al reported 114 (48%) cases of aspiration syndrome due to neurodevelopmental delay in a series of 220 cases of aspiration pneumonias.[12]

Our study underlines the importance of oropharyngeal incoordination in causing respiratory complications in children with cerebral palsy. In a study it was found that the presence of direct aspiration to be the best marker of defective oropharyngeal function and was considered preferable than a detailed analysis of the oral and pharyngeal stages of swallowing.[21] Though now a days video fluoroscopic swallow study (VFSS) for evaluation of swallowing problems is method of choice rather it is prophylactically recommended in all children with neurodevelopmental abnormalities.[8,24,25,26] however, in our set up due to non availability of the VFSS our diagnosis was based upon clinical and radiological findings. When considering the diagnosis of recurrent aspiration observation of feeding is essentially recommended with particular attention being paid to nasopharyngeal reflux, gag reflex, difficulty while sucking or swallowing and associated coughing, gagging and choking. Presence of drooling or excessive accumulation of secretions in the mouth on examination of oral cavities suggest dysphagia with resultant chronic aspiration.[18]

In our study gastroesophageal reflux was clinically suspected in quite a number of cases, however, it was difficult to pin point the exact number of cases with significant gastroesophageal reflux disease due to the lack of 24hrs lower esophageal pH monitoring. It has been noted that the presence of gastroesophageal reflux makes these neurodevelopmentally delayed children more prone not only to indirect aspiration but also to even direct aspiration because of higher chances of aspiration in the presence of chronic inflammation of the pharynx resulting from the noxious effects of refluxate.[21,22] The pneumonia in children having GER and cerebral palsy both is more severe because it is likely that they aspirated the acidic refluxate from the stomach, which is particularly noxious to the lungs.[21,23]

Our study has certain limitations. (i) The radiological diagnosis of pneumonia in our study is susceptible to bias. As differentiation between atelectasis and consolidation is not always possible from x-ray chest, the diagnosis of pneumonia might have been overestimated.(ii) The direct demonstration of aspiration into airways was not possible because of the non-availability of Video fluoroscopic swallowing studies (VFSS), so the evidence of aspiration was indirect i.e. clinical plus radiological. (iii) Moreover, due to different financial and geographical constraint certain tests like sweat chloride test and complete immunological workup could not be performed, we might have missed few associated causes of recurrent pneumonia other than cerebral palsy.

CONCLUSION

Cerebral palsy is a common disorder leading to significant disability and deaths. Respiratory problems especially recurrent pneumonia is a common complication of cerebral palsy. Solid and sustainable strategies need to be developed and implemented to reduce the incidence and complications of cerebral palsy. The major issues desiring urgent attention include improvement in maternal health, antenatal and perinatal services along with neonatal care.

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