Original Articles

Pulmonary Function and Chest Expansion in Thai Boys with Down Syndrome

Sirinart Laibsirinon, Noppawan Jarusurin,
Chalanda Kokoi, Titarat Manakiatichai

Abstract

Pulmonary problems in children with Down syndrome may contribute to decreased level of physical fitness, compared to healthy children. Pulmonary function in children with Down syndrome has been little known. Inadequate amount of oxygen or air volume during inspiration and expiration measured by using a handheld spirometer and limitation of chest expansion may be used to indicate the problems of pulmonary function in children with Down syndrome. Force expiratory volume in a second (FEV1) and chest expansion of 10 boys with Down syndrome was significantly different from those of 10 healthy boys. The trend of force vital volume capacity (FVC) was also reduced in boys with Down syndrome. These findings might be the result of weakness of primary and accessory respiratory muscles, particularly abdominal muscles, and narrow respiratory airway. Medical scientists and health-related staffs who work with children with Down syndrome should also focus on improving their respiratory status and preventing possible pulmonary infection. Pulmonary function and chest expansion test have potential to be an initial indicator of pulmonary problems in people with Down syndrome at a young age.

Key words: Down syndrome, Pulmonary function, Chest expansion

Introduction

Pulmonary problems in children with Down syndrome are under-recognized and may partly hinder a child's ability to achieve optimal level of physical activity and fitness. Low level of physical fitness is a major issue for children with Down syndrome.[1,2] Persistent poor physical fitness from childhood through adulthood will increase higher risks for health-related issues such as osteoporosis, type 2 diabetes, obesity, and cardiopulmonary diseases.[3-5] Physical fitness is defined as the ability to perform daily activities without fatigue. It incorporates the characteristics of body composition, flexibility, muscular strength and endurance, and cardiovascular fitness.[6] These five characteristics have long been the major area of research interest, and are used to identify level of physical fitness in people, including children with Down syndrome.[7-11] In contrast, there are only a few published researches investigating pulmonary function in children with Down syndrome.[12,13] Children with Down syndrome have been reported with upper and lower airway abnormalities including a small upper airway, decreased numbers of alveoli and reduced surface area.[14-16] Generalized weakness, especially abdominal muscle, is prominent in people with Down syndrome patients.[17] These two major problems can lead to decreased lung volume. Cardiovascular and respiratory systems work closely together to ensure sufficient amount of oxygen carries to organs. If lung volume or amount of maximum oxygen is reduced, removal of carbon dioxide is deficient, therefore limiting available energy for activities. Pulmonary problems of children with Down syndrome may be a key factor that leads to poor level of physical fitness.

Department of Physical Therapy, Faculty of Allied Health Sciences, Thammasat University

Although pulmonary function norms have been established in healthy children, it is the norms of Caucasian children.[18,19] These norms may not be applicable to Thais because of ethnicity, lifestyle, and cultural differences. Chest expansion, which has the potential to be used as an early indicator for detecting pulmonary problems, has never been studied in children with Down syndrome. Pulmonary function tests and chest expansion are simple and quantitative measurement that can be used to determine a baseline respiratory function in place of inclusive and expensive diagnostic methods such as chest x-ray, computerized topography (CT) and bronchoscopy. Purposes of this study were: 1) to evaluate pulmonary function and chest expansion in children with Down syndrome; 2) to compare the findings with those of healthy children.

Materials and Methods

Subjects

Ten boys with Down syndrome and ten healthy boys participated in this study. They ranged in age from 9 to 12 years old (mean ± SD: 10.0 ± 1.1 years in Down syndrome and 10.2 ± 0.6 years in healthy boys). Height and body mass index (BMI) of healthy boys were matched with Down syndrome group. Inclusion criteria of both groups included: 1) no spinal abnormality such as kyphosis and scoliosis; 2) no history of pulmonary infection and surgery of thoracic and abdominal region within the past 6 months. Both groups of children performed typical daily activities and did not play any specific sport or exercises. Boys with Down syndrome attended a special private school providing educational program for students with intellectual disability. They did not have any current cardiac symptom or disorders as reported in their medical records. All boys with Down syndrome were classified as moderate mental retardation, which meant they could follow commands and had no physical activity restrictions, as cited in their school records. Legal guardian or a parent signed consent forms prior to each subject's participation. This study was approved by the Thammasat University Ethics Subcommittee.

Equipments and test procedures

There were two tests consisting of pulmonary function test and chest expansion. Prior to data collection, subjects from each group were instructed with equipment and explained about test procedures. Practice sessions were provided to ensure that the subjects understood and were able to perform the tests. They could practice the tests until they were familiar with the equipments and test procedures. There was a 10-minute rest between pulmonary function and chest expansion test in order to prevent the subject from exhaustion and fatigue.

1. Pulmonary function test

Pulmonary function was measured using a hand-held spirometer (Micro Medical Company, Kent, UK). The subject sat erect on a chair with feet flat on the ground. The subject's nose was clipped with a soft nose clip. A paper mouth piece connected to the tubing was placed in the mouth. While performing the test, lip closure around the mount piece had to be secured. The subject was asked to take the deepest breath, and then exhaled as quickly and as hard as possible. A researcher coached the subject whereas the other recorded the test result. The spirometer was clean, disinfected and reset to zero each time for each of the subjects. Three trials with at least a 2-minute rest were collected. The best values among three trials of force vital capacity (FVC) and forced expiratory volume in 1 second (FEV_1) recorded in liters were used for statistical analysis.

2. Chest expansion

Chest expansion is defined as chest wall mobility or excursion between a maximum voluntary inspiration and a maximum voluntary expiration. The circumference of chest wall was measured using a tape measure. Moderate to high intra-tester reliability of measurement of chest expansion with a use of a tape measure was reported.[20] This test was taken when the subject sat straight on the chair. A tape measure was placed at the level of the 7^{th} intercostal space or at the level of inferior aspect of the xiphoid process. The xiphoid process had been standardized as an appropriate level for lower chest wall excursion

which possibly might be the most problematic in children with Down syndrome.[21, 22] While a researcher instructed the subject to take maximum inhalation and continue to exhale completely, another researcher performed the test and recorded the results in centimeters. Three trials were performed with at least a 2-minute rest. Among results from the three trials, the greatest difference of chest circumference between maximum inspiration and expiration was used in the statistical analysis.

Data analysis

The normality of the data was tested using Kolmogorov-Smirnov Goodness of Fit test. Independent Student's t test was performed to find the difference of subject characteristics between boys with Down syndrome and healthy boys. Because of a small sample size and non-normality distribution of the data, the Mann-Whitney U test was used to find the differences of FVC, FEV_1 and chest expansion between the two groups. The level of significance was set at 0.05. SPSS version 17.0 software (SPSS, Chicago, IL) was used for all analyses.

Results

Weight, height, and BMI between boys with Down syndrome and healthy boys were not significantly different ($p>0.05$). Descriptive statistics of subject's characteristic are shown in table 1.

Table 1 Characteristics of boys with Down syndrome and healthy boys

Characteristics	*Down syndrome (n = 10)*	*Healthy boys (n = 10)*	*p-value*
Age (year)	10.00 ± 1.0	10.2 ± 0.6	0.61
Weight (kg)	37.30 ± 5.33	38.30 ± 2.83	0.61
Height (cm)	1.29 ± 0.10	1.26 ± 0.03	0.45
BMI (kg/m^2)	22.47 ± 2.56	23.97 ± 1.77	0.14

Table 2 Chest expansion and pulmonary function of boys with Down syndrome and healthy boys

Variables	*Down syndrome (n = 10)*	*Healthy boys (n = 10)*	*p-value*
FVC (liter)	1.45 ± 0.75	1.77 ± 0.25	0.22
FEV_1 (liter)	1.06 ± 0.47	1.60 ± 0.20	0.01*
Chest expansion (cm)	3.50 ± 0.86	4.81 ± 1.25	0.02*

* Statistically significant difference, at $p>0.05$

Table 2 presents descriptive statistics for the result from pulmonary function test and chest expansion. There was no statistically significant difference of force vital capacity (FVC) between boys with Down syndrome and healthy boys ($p=0.22$) whereas FEV_1 was significantly different ($p=0.01$). Values of FVC and FEV_1 of boy with Down syndrome were lower than those of healthy boys.

Mean and standard deviation of chest expansion of Down syndrome and healthy boys was 3.50 ± 0.86 and 4.81 ± 1.25 cm, respectively. A significant difference of chest expansion was found between these two groups.

Discussion

Any significant result found in this study did not attribute to variations in size, gender and age of the subject between boys with Down syndrome and healthy boys since the age, height, and BMI were not significantly

different. The findings should not be an error in lack of understanding of children with Down syndrome or lack of cooperation from both groups of children. They all were highly motivated and showed active participation in the tests. Children with Down syndrome participating in this study were also students at a special school. They cooperated and could follow the command and instruction very well. The intensive practices of chest expansion test and spirometry use were also provided for both groups. Thus, the significant differences of lung function and chest expansion obtained in this study should be the actual findings between boys with Down syndrome and healthy boys.

The values of FVC and FEV_1 in children with Down syndrome obtained from this study were higher than those from Dichter's study.[12] This may contribute to variation in result from differences in gender of all participants between 2 studies. Only boys with Down syndrome participated in this study whereas there were 6 girls out of 18 children with Down syndrome in the study of Dichter et al. It has been reported that FVC and FEV_1 are influenced by gender differences because boys have larger lungs and narrower airways than girls.[23] Differences in ethnic origin and height may also contribute to this disagreement.[19,24,25] However, this is not conclusive because mean height of all participants in Dichter's study were not reported. Furthermore, it might be due to the different types of spirometer used between 2 studies. A water-sealed bell spirometer connected with a microcomputer was used in the study of Dichter whereas a handheld spirometer with a digital reading was used in this study. A handheld spirometer is a portable lightweight device and is easily held by one hand which may be more convenient for the participants. Children may feel less nervous when performing the test with a handheld spirometer.

A significantly lowered FEV_1 and a tendency toward lowered FVC in boys with Down syndrome, compared to healthy boys indicate problems with pulmonary system. The contributing factors of pulmonary problems may include muscle weakness, particularly abdominal muscle and smaller upper airway size in children with Down syndrome.[9,15,17,26] During normal quiet breathing, ventilation or adequate air volume is achieved by a contraction of diaphragm and the external intercostal muscle during inspiration and a passive process of recoil of the chest wall during expiration. During forced breathing, the accessory muscles of neck and shoulder assist in forced inspiration while abdominal muscle is primarily responsible for forced expiration.[27] Although weakness of neck and shoulder muscles of Down syndrome has never been published, it has been noted and recognized in clinics.[28] Forwarded neck posture, which is assumed to be caused by weakness of neck muscle, is a common sitting position for children with Down syndrome. Weakness of abdominal muscle has been widely reported and is critical in poor coughing in children with Down syndrome.[28,29] Effective cough is in linear relation to force expiration volume. Weak cough or lowered FVC necessitates high incidences of mucous accumulation that results in respiratory infection. High incidences of respiratory infection are common in children with Down syndrome.[30,31] Smaller upper airway size is presented by MRI examination in children with Down syndrome, compared to healthy children of the given age.[15] Smaller airway increased resistance of airflow during breathing. As such, weakness of respiratory muscles and narrow airway size contribute to a reduction of airflow, which in turn, lead to inadequacy of oxygen or energy to perform normal physical activities. Reduced physical activity and poor physical fitness in children with Down syndrome, compared to healthy children are commonly reported.[1,2]

Chest expansion of boys with Down syndrome was significantly smaller than healthy boys. Chest expansion or mobility of chest wall is affected by normal function of respiratory muscles, and mobility of costoverterbral joints as well as compliance of lung. Unfortunately, these three factors have never been thoroughly examined in Down syndrome. If most people with Down syndrome have no difficulty in normal breathing and have normal breathing pattern, it is reasonable to believe that lowered chest expansion of Down syndrome may be partially due to weakness of respiratory muscles (inspiratory and expira-

tory muscles). However, this conclusion needs further investigation.

Medical personnel working with children with Down syndrome should also put emphasis on their pulmonary function. Early initiation of rehabilitation or exercise program should be provided to improve and maintain pulmonary function in children with Down syndrome. The improvement of pulmonary function after an 8-week program of aerobic exercise was found in children with intellectual disability, including Down syndrome.[26] In addition to cardiovascular exercise, physical fitness program should include strengthening and endurance exercise for respiratory muscle and intensive strengthening exercise for abdominal muscles. Primary and accessory respiratory muscles may be exercised by using an incentive spirometer. Hydrotherapy may be beneficial to children with Down syndrome. Furthermore, chest expansion and pulmonary function test are feasible, simple and inexpensive methods that may be used in clinical setting in order to screen baseline status of pulmonary function, to detect pulmonary problems early and to monitor gain from physical fitness program.

Conclusion

Pulmonary function (FVC and FEV_1) and chest expansion in boys with Down syndrome were lower than healthy boys. It indicates pulmonary problems which may result from weakness of accessory respiratory muscle, abdominal muscle and smaller size of upper airway. Generalization of this study should be cautious due to the small sample size. Further research is required to determine the relationship of pulmonary function and strength of abdominal muscle in children with Down syndrome.

References

1. Fernhall B, Pitetti KH, Rimmer JH, McCubbin JA, Rintala P, Millar AL, et al. Cardiorespiratory capacity of individuals with mental retardation including Down syndrome. Med Sci Sports Exerc 1996;28:366-71.
2. Whitt-Glover MC, O'Neill KL, Stettler N. Physical activity patterns in children with and without Down syndrome Pediatr Rehabll 2006;9:158-64.
3. Prasher VP. Overweight and obesity amongst Down's syndrome adults. J Intellect Disabil Res 1995;39:437-41.
4. Draheim CC, Williams DP, McCubbin JA. Prevalence of physical Inactivity and recommended physical activity in community-based adults with mental retardation. Men Retard 2002;40:436-44.
5. Barnhart RC, Connolly B. Aging and Down Syndrome: Implications for Physical Therapy. Phy Ther 2007;87:1399-406.
6. Pollock ML, Gaesser GA, Butcher JD, Després J-P, Dishman RK, Franklin BA, et al. American College of Sports Medicine Position Stand: The Recommended Quantity and Quality of Exercise for Developing and Maintaining Cardiorespiratory and Muscular Fitness, and Flexibility in Healthy Adults. Med Sci Sports Exerc 1998;30:975-91.
7. Luke A, Roizen NJ, Sutton M, Schoeller DA. Energy expenditure in children with Down syndrome: Correcting metabolic rate for movement. J Pediatr 1994;125:829-38.
8. Angelopoulou N, Matziari C, Tsimaras V, Sakadamis A, Souftas V, Mandroukas K. Bone Mineral Density and Muscle Strength in Young Men with Mental Retardation (With and Without Down Syndrome). Calci Tissue Int 2000;66:176-80.
9. Mercer VS, Lewis CL. Hip abductor and knee extensor muscle strength of children with and without Down syndrome. Peditr Phys Ther 2001;13:18-26.

10. Carmeli E, Barchad S, Lenger R, Coleman R. Muscle power, locomotor performance and flexibility in aging mentally-retarded adults with and without Down's syndrome. J Musculoskel Neuron Interact 2002;2: 457-62.
11. Dodd KJ, Shields N. A Systematic Review of the Outcomes of Cardiovascular Exercise Programs for People With Down Syndrome. Arch Phy Med Rehabil 2005;86:2051-8.
12. Dichter CG, Darbee JC, Effgen SK, Palisano R. Assessment of pulmonary function and physical fitness in children with Down syndrome. Peditr Phys Ther 1993;5:3-8.
13. Pastore E, Marino B, Calzolari A, Digilio MC, Giannotti A, Turchetta A. Clinical and cardiorespiratory assessment in children with Down Syndrome without congenital heart disease. Arch Pediatr Adolesc Med 2000;154:408-10.
14. Schloo BL, Vawter GF, Reid LM. Down syndrome: Patterns of disturbed lung growth. Hum Patho 1991;22:919-23.
15. Uong E, McDonough J, Tayag-Kier C, Zhao H, Haselgrove J, Mahboubi S, et al. Magnetic resonance imaging of the upper airway in children with Down syndrome. Am J Respir Crit Care Med 2001;163:731-6.
16. McDowell K, Craven D. Pulmonary complications of Down syndrome during childhood. J Pediatr 2011;158:319-25.
17. Harris S, Shea A. Down syndrome. In: Campbell S, editor. Pediatric neurologic physical therapy. 2nd ed. New York: Churchill Livingstone; 1991. p. 131-68.
18. Bernstein IL, Fragge RG, Gueron M, Kreindler L, Ghory JE. Pulmonary function in children: I. Determination of norms. J Allergy 1959;30:514-33.
19. Wang X, Dockery DW, Wypij D, Fay ME, Ferris BG. Pulmonary function between 6 and 18 years of age. Pediatr Pulmono 1993;15:75-88.
20. Stackowicz D, Frownfelter D, Rheault W. Intertester reliability of the measurement of chest wall expansion. Cardiopulmonary Phy Therapy J 2003;14:24.
21. Bockenhauer SE, Chen H, Julliard KN, Weedon J. Measuring Thoracic Excursion: Reliability of the Cloth Tape Measure Technique. J Am Osteopath Assoc 2007;107:191-6.
22. Malaguti C, Rondelli RR, de Souza LM, Domingues M, Dal Corso S. Reliability of Chest Wall Mobility and Its Correlation With Pulmonary Function in Patients With Chronic Obstructive Pulmonary Disease. Respir Care 2009;54:1703-11.
23. Carey MA, Card JW, Voltz JW, Arbes Jr SJ, Germolec DR, Korach KS, et al. It's all about sex: gender, lung development and lung disease. Trends Endocrinol Metab 2007;18:308-13.
24. Rossiter CE, Weill H. Ethnic Differences in Lung Function: evidence for proportional differences. Int J Epidemiol 1974;3:55-61.
25. Whittaker A, Sutton A, Beardsmore C. Are ethnic differences in lung function explained by chest size? Arch Dis Child Fetal Neonatal Ed 2005;90:423-8.
26. Khalili MA, Elkins MR. Aerobic exercise improves lung function in children with intellectual disability: a randomised control trial. Aust J Physiother 2009;55:171-5.
27. Dias K. Physiology of the cardiovacular systems. In: Hillegass E, editor. Essential of cardiopulmonary physical therapy. St.Louis: Elsevier Saunders; 2011. p. 27-46.
28. Bergeron K, Dichter CG. Case study: Down syndrome. In: Effgen SK, editor. Meeting the physical therapy needs of children. Philadelphia: F.A. Davis company; 2005. p. 516-38.
29. Lauteslager PEM, Vermeer A, Helders PJM. Disturbances in the Motor Behaviour of Children with Down's Syndrome: The need for a theoretical framework. Physiother 1998;84:5-13.

30. Yang Q, Rasmussen SA, Friedman JM. Mortality associated with Down's syndrome in the USA from 1983 to 1997: a population-based study. Lancet 2002;359:1019-25.

31. Trotsenburg ASPv, Heymans HSA, Tijssen JGP, Vijlder JJMd, Vulsma T. Comorbidity, Hospitalization, and Medication Use and Their Influence on Mental and Motor Development of Young Infants With Down Syndrome. Pediatrics 2006;118:1633-9.

บทคัดย่อ

การทำงานของปอด และการขยายตัวของทรวงอกในเด็กชายไทยที่มีภาวะดาวน์ซินโดรม

ศิรินาถ เลียบศิรินนท์, นพวรรณ จารุสุสินธ์, ชลันดา โก๊ะกอย, ฐิตารัตน์ มานะเกียรติชัย
ภาควิชากายภาพบำบัด คณะสหเวชศาสตร์ มหาวิทยาลัยธรรมศาสตร์

ปัญหาทางด้านปอดในเด็กที่มีภาวะดาวน์ซินโดรม อาจจะส่งผลให้เด็กกลุ่มนี้มีสมรรถภาพทางกายที่ลดลงเมื่อเปรียบเทียบกับเด็กที่มีสุขภาพดีทั่วไป จากการทบทวนวรรณกรรมที่ผ่านมาพบว่า มีการศึกษาถึงการทำงานของปอดในเด็กที่มีภาวะดาวน์ซินโดรมน้อยมาก ปัญหาของการทำงานของปอด (pulmonary function) ในเด็กที่ภาวะดาวน์ซินโดรม อาจจะสามารถบ่งบอกได้จากปริมาณของออกซิเจนที่ไม่เพียงพอในขณะที่หายใจเข้าและหายใจออก ซึ่งจะสามารถวัดได้โดยการใช้เครื่องอินส์ไปโรมิเตอร์แบบพกพา และสามารถบ่งบอกได้จากการจำกัดการขยายตัวของทรวงอก (chest expansion) จากการศึกษา พบว่า ปริมาณอากาศในช่วงหายใจออกอย่างแรง ใน ๑ วินาที (Force expiration volume in a second) และค่าของการขยายตัวของทรวงอกมีความแตกต่างอย่างมีนัยสำคัญระหว่างเด็กชายไทย ๑๐ รายที่มีภาวะดาวน์ซินโดรมและเด็กชายไทย ๑๐ รายที่มีสุขภาพดีทั่วไป นอกจากนั้นยังพบแนวโน้มของการลดลงของค่าความจุชีพ จากการเป่าอย่างแรง (Force vital capacity) ในเด็กชายไทยที่มีภาวะดาวน์ซินโดรม ทั้งนี้อาจเกิดขึ้นเนื่องจากในกลุ่มเด็กชายไทยที่มีภาวะดาวน์ซินโดรม อาจจะมีการอ่อนแรงของกล้ามเนื้อหลักและเสริมที่ใช้ในการหายใจ โดยเฉพาะกล้ามเนื้อหน้าท้อง และการตีบแคบของช่องทางเดินหายใจ บุคลากรทางการแพทย์และผู้ที่ทำงานเกี่ยวข้องกับเด็กที่มีภาวะดาวน์ซินโดรม น่าจะส่งเสริมการพัฒนาศักยภาพการทำงานของปอด และป้องกันการเกิดติดเชื้อทางระบบหายใจที่พบบ่อยในเด็กที่มีภาวะดาวน์ซินโดรม จากผลการศึกษาครั้งนี้พบว่าการทดสอบสมรรถภาพการทำงานของปอดและการขยายตัวของทรวงอก อาจนำมาใช้ทางเวชกรรมเพื่อบ่งบอกถึงระดับของสมรรถภาพของปอดหรือเป็นตัวชี้วัดเบื้องต้นที่บ่งบอกถึงการเกิดปัญหาของการติดเชื้อที่ปอดในเด็กที่มีภาวะดาวน์ซินโดรมได้

คำสำคัญ: ภาวะดาวน์ซินโดรม, การทำงานของปอด, การขยายตัวของทรวงอก