Chapter 13
Case-Based Analysis of Respiratory Failure

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As described so far, respiratory failure and respiratory distress have varied etiologies and each case has to be analyzed, diagnosed, investigated and managed individually. While certain broad principles are common to groups of disorders, each patient has his own unique features in presenting symptoms and signs, clinical course, and response to treatment. We present a systematic approach to the analysis of symptomatology, clinical manifestations and pathophysiologic considerations in the diagnosis and management of respiratory failure of varied etiology.

13.1 Breathing Control Disorders

Case 1

A 14-year-old boy presented to the emergency room with somnolence, lethargy, and difficulty in arousing. For the past month, he had been complaining of tiredness, neck pain, and early morning headaches. He was afebrile with shallow respirations of 10/min, heart rate of 88/min and blood pressure of 160/100 mmHg. Pulse oximetry showed SpO_2 of 90%. Pupils were 4 mm and reacted briskly to light. No evidence of

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trauma was noted. He responded to external stimulation and answered simple questions but went back to sleep. The rest of the physical examination including that of chest and abdomen was normal. Arterial blood gas in room air revealed pH 7.21, PCO_2 70 torr and PO_2 62 torr. Supplemental O_2 via a face mask was applied. Fifteen minutes later, he was found to be unarousable with barely discernible respiratory effort. SpO_2 was 100%. An arterial blood gas showed pH 7.0, PCO_2 100 torr and PO_2 120 torr. Rapid sequence intubation was performed, and volume-controlled ventilation was instituted with respiratory rate 18/min, tidal volume (V_T) 500 ml, inspiratory time (T_i) 1 s and PEEP of 4 cm H_2O. Chest radiograph revealed normal lungs and heart and endotracheal tube at mid-tracheal level.

Clinical Analysis

The patient has severe respiratory failure however, he shows no signs of respiratory distress, has no abnormal physical findings except for weak respiratory effort. His chest radiograph also shows no abnormal lungs or heart findings. Gas exchange shows severe hypoventilation without significant oxygenation deficit suggesting that this is an abnormality of alveolar ventilation with sparing of alveolar capillary apparatus. Central (above the carina) airway obstruction at the level of pharynx, larynx or trachea could explain alveolar hypoventilation. However, lack of respiratory effort to overcome airway obstruction severe enough to cause this level of respiratory failure, makes this group of disorders unlikely. This leaves us with two possibilities: a) ineffective neuro-musculoskeletal function or b) abnormal central control of respiration. Patients with neuropathies (Guillain Barre syndrome, cervical spinal cord injuries etc.), myopathies (muscular dystrophies, botulism etc.) and skeletal abnormalities have diminished capacity to mount effective effort to combat hypercarbia. However, such patients are conscious and responsive. Somnolence and lack of arousal observed in this patient make this possibility also unlikely. That leaves us with the abnormal central control of respiration as the most likely possibility.

By far the most common cause of decreased central chemoreceptor responsiveness is exposure to CNS sedatives such as drugs of abuse or therapeutic administration. Other forms of toxic, infectious, traumatic, and metabolic encephalopathies could also lead to decreased central chemoreceptor responsiveness. The observation that this patient developed further depression of respiration after administration of supplemental O_2 suggests that his control of respiration was much more dependent on hypoxia sensing peripheral chemoreceptors rather than CO_2 sensing central chemoreceptors. Such a situation occurs in brainstem lesions as well as chronic CO_2 retention where central chemoreceptors are blunted or downregulated. Once peripheral chemoreceptor stimulation was abolished in this patient by supplemental oxygen and a rise in PaO_2, the respiratory drive was diminished resulting in CO_2 narcosis due to ineffective central chemoreceptors.

Subsequent Course

After mechanical ventilation and decreasing $PaCO_2$ to normal levels, and as the effect of pre-intubation drugs subsided, the patient awoke and was interactive. He was extubated and a nasal cannula delivering supplemental O_2 was applied. Several

hours later he became somnolent again with development of respiratory acidosis requiring reintubation. The clinicians recognized that this was a disorder of central control of respiration and that the patient was mainly dependent on the hypoxic stimulation of peripheral chemoreceptors for spontaneous respiration. Abolishing hypoxic stimulation with supplemental O_2 was blunting his ventilatory drive resulting in hypercarbia and CO_2 narcosis. Brain and brain stem magnetic resonance imaging (MRI) was performed revealing Type I Chiari malformation. Patient underwent sub occipital craniotomy and decompression of the posterior fossa relieving the pressure on his brainstem. Responsiveness of central chemoreceptors to CO_2 was restored and the patient was discharged home with full recovery.

Important Points

1. Pure alveolar hypoventilation results from central airway obstruction, depressed respiratory center, and ineffective neuromuscular function. Clinical manifestations provide clues to diagnosis.
2. Central chemoreceptors are responsive to acute changes in PCO_2 by decreasing the CSF pH. Peripheral chemoreceptors are primarily responsive to hypoxia, but also to hypercarbia to some extent.
3. Response time to peripheral chemoreceptor stimulation is much more rapid than that of central chemoreceptor stimulation.
4. The magnitude of response to acutely elevated PCO_2 is much greater via stimulation of central chemoreceptors compared to peripheral chemoreceptors
5. Peripheral chemoreceptor stimulation persists over a long time, even for a life-time. Central chemoreceptors however get acclimatized over time with blunting of response to elevated $PaCO_2$. In such situations, removal of peripheral chemoreceptor stimulation by O_2 administration could result in serious hypoventilation and CO_2 narcosis.

13.2 Heart Failure

Case 2

A three-month-old infant boy with Down's syndrome and atrioventricular (AV) canal was brought to the emergency department for increasing respiratory difficulty, grunting and retractions for 12 h. Temperature is 38.4 °C, respirations of 36/min, heart rate of 140/min and blood pressure of 90/45 mmHg. Precordial heave is noted. Chest auscultation revealed grade IV holosystolic murmur along the left sternal border. Expiratory wheezing and grunting were heard. SpO_2 was 90%. Chest radiograph showed cardiomegaly, with prominent central vascular markings and air trapping. High flow nasal cannula was applied with 40% O_2. Thirty minutes later, the infant was noted to be diaphoretic with respiratory rate of 60/min, heart rate of 160/min, blood pressure of 88/58 mmHg and SpO_2 of 100%. Pulses were

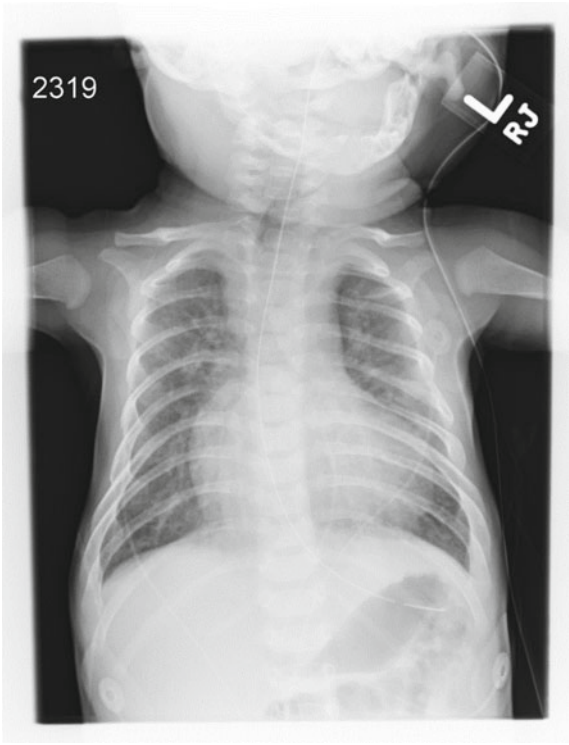

Fig. 13.1 Chest radiograph of a three-month-old male infant shows moderate cardiomegaly, pulmonary atrial and venous congestion, and hyperinflation of lungs

noted to be thready; peripheries were cool with delayed capillary refill. Repeat chest radiograph (Fig. 13.1) showed worsening of cardiomegaly and pulmonary edema.

Clinical Analysis

This infant has a large left-to-right shunt. The systemic arterial blood flow (Q_s) is diverted to the pulmonary arterial flow (Q_p) at both atrial and ventricular level. Such is the case with large ventricular septal defect, atrial septal defect, patent ductus arteriosus and aortopulmonary window. In a normal heart, without any communication between right-sided and left-sided circulations, Q_p is equal to Q_s. In left-to-right shunts, Q_p is greater than Q_s because of lower pulmonary vascular resistance (PVR) compared to the systemic vascular resistance (SVR) The extent to which Q_p exceeds Q_s depends on the size of the shunt and the respective resistances in systemic and pulmonary circulations. The PVR remains fairly high in the neonatal period limiting the left-to-right shunt. As the infant grows, the PVR steadily declines until at the age of 3–6 months, when it reaches the adult value of being 15–20% of SVR. With significantly increased Q_p, the pulmonary blood volume and the interstitial fluid are increased, resulting in decreased lung compliance. Baseline respiratory rates tend to be increased in such situations. The infant probably had respiratory decompensation with an intercurrent viral infection and

presented with respiratory distress. Supplemental O_2 administration resulted in a further decrease in PVR increasing the left-to-right shunt, increased Q_p:Q_s ratio and worsening of pulmonary congestion and edema. While SpO_2 increased with greater Q_p, he developed systemic hypoperfusion as flow was diverted to the lung from the rest of the body. Maintaining SpO_2 in low 90's should be sufficient for oxygenation without compromising systemic perfusion. Mechanical support of respiration should be aimed at improving FRC and using a low V_T strategy since we are dealing with a disease of compliance. The amount of FiO_2 delivered should be enough to maintain SpO_2 in the low 90's since higher SpO_2 carries the risk of decreasing PVR and increasing the Q_p:Q_s ratio. Use of optimum PEEP (along with positive pressure breathing) will be very helpful since it will: (a) displace alveolar fluid to the extra-alveolar space allowing for better gas exchange thereby reducing the FiO_2 requirement, (b) improving compliance by decreasing the pulmonary blood volume thereby allowing lower inflation pressure, (c) increasing the right ventricular afterload by alveolar capillary compression and therefore decreasing the left-to-right shunt and (d) decreasing left ventricular afterload by positive intrathoracic pressure during both the isovolumic contraction and the ejection phases of systole. The disadvantages of invasive mechanical ventilation in this situation are primarily related to peri-intubation stress and trauma and the use of the necessary sedatives/paralytic agents. Intubation should be performed in controlled conditions.

Subsequent Course

The infant was intubated after appropriate sedation and pharmacologic paralysis. Pressure-regulated volume control (PRVC) ventilation was instituted to deliver 7 mL/kg V_T with a peak inspiratory pressure (PIP) reaching 25 cm H_2O, inspiratory time of 0.8 s, PEEP of 6 cm H_2O, ventilator rate of 25/min and FiO_2 of 0.3. Furosemide was administered for diuresis. Subsequently, the patient exhibited improved perfusion. Over the next 36 h, mechanical ventilation was weaned off and the patient was successfully extubated.

Important Points

1. O_2 administration may result in pulmonary vasodilation in patients with large left-to-right shunts (e.g. VSD, AV canal) and functionally univentricular lesions (e.g. after Norwood operation). In such situations Qp:Qs may increase to such an extent as to result in pulmonary vascular overload, pulmonary edema and systemic hypoperfusion.
2. Positive pressure ventilation is an effective means of decreasing LV afterload. In patients with poor ventricular contractility (e.g. myocarditis, cardiomyopathy), positive pressure ventilation is an effective strategy to mechanically decrease the LV afterload and improve cardiac output.

13.3 Lower Airway Obstruction

Case 3

A 12-year-old boy presents to the emergency department with exacerbation of his known asthma and increasing difficulty breathing for the last 8 h. He is unable to speak in complete sentences. Vital signs are temperature of 37.8 °C, respirations of 24/min, heart rate of 110/min and blood pressure of 108/72 mmHg with pulsus paradoxus of 40 mmHg. SpO_2 while breathing 40% O_2 via a Venturi mask is 89%. Extremities are cool with dusky nailbeds. Examination reveals marked intercostal and subcostal retractions, prolonged expiratory phase and audible wheezing. Very little response to inhaled albuterol and intravenous steroids is noted. Arterial blood gases show pH of 7.12, PCO_2 of 105 torr and PO_2 of 70 torr on non-rebreather O_2 mask. The patient appears to be tiring and lethargic. Rapid sequence intubation is performed and arrangements are made to transport to the ICU for further management. Chest radiograph shows bilateral hyperinflation, flattened diaphragms, perihilar infiltrates, prominent main pulmonary artery, and a relatively small cardiac silhouette (Fig. 13.2). No air-leak is noted. Patient is being manually ventilated via a bag to ET tube with 100% O_2 at a rate of 30/min and inflation pressure of 50 cm H_2O. Repeat arterial blood gas analysis shows a pH of 6.95, PCO_2 of 130 torr and PO_2 of 104 torr.

Clinical Analysis

This patient has marked intrapulmonary airway obstruction with non-uniform ventilation and severe V/Q mismatch. His respiratory time constant (compliance X

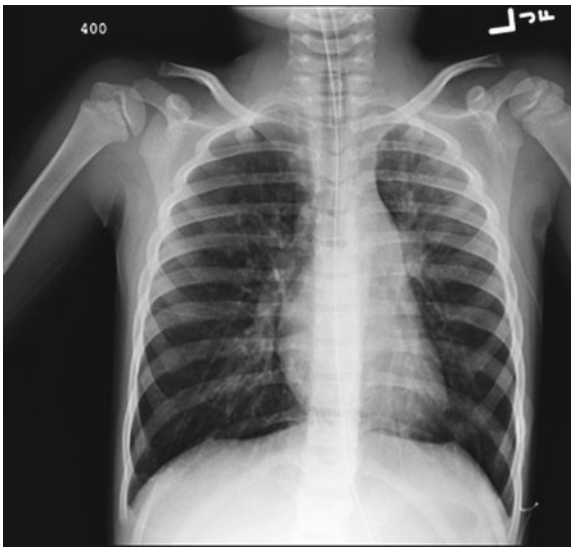

Fig. 13.2 Chest radiograph of a 12-year-old boy with status asthmaticus. Note severe bilateral lung hyperinflation, flattened diaphragm and prominent pulmonary artery. Perihilar infiltrates are seen bilaterally

resistance) is markedly prolonged and since the obstruction is intrapulmonary, his equal pressure point (EPP) is displaced far more distally resulting in widespread airway collapse during exhalation. Thus, the expiratory time constant is much more prolonged compared to the inspiratory time constant which is also prolonged. With insufficient time for complete exhalation, there is air-trapping, auto-PEEP, decreased dynamic compliance, impaired venous return and decreased cardiac output. Markedly increased pulsus paradoxus before intubation is indicative of wide swings in intrathoracic pressure during inspiration and exhalation, decreased LV preload and increased LV afterload during inspiration. In order to deliver tidal volume effectively, we need to use sufficient time for both inspiration and exhalation because of his prolonged time constants. This can only be achieved by decreasing the respiratory rate to allow for longer time for each respiratory cycle. Furthermore, exhalation time (T_e) should be more prolonged compared to inspiratory time (T_i) to allow for greater alveolar emptying. This would improve dynamic compliance, decrease auto-PEEP, reduce the risk of barotrauma and increase venous return. Thus, the strategy should be that of a low mandatory rate with a high PIP/or V_T ventilation while monitoring both exhaled V_T and alveolar ventilation. His gas exchange worsened after intubation and manual ventilation. This is most likely because of being ventilated at too high a rate to allow for adequate delivery of V_T and alveolar emptying. On mechanical ventilation, PRVC or pressure controlled ventilation (PCV) would allow for better distribution of delivered volume compared to volume controlled ventilation which will be preferentially distributed to relatively lower resistance areas compared to higher resistance areas with resultant increased inflation pressure and barotrauma. A certain amount of PEEP would help to move EPP more proximally, decrease transmural pressure and reduce airway collapse. Inspiratory and expiratory flows along with auto-PEEP should be monitored by flow-time waveforms. Similarly, exhaled tidal volume should be monitored at different ventilator rates and PIP/V_T combinations to deliver a desired minute ventilation with minimum injury. The threat to life in status asthmaticus is mainly from hypoxia and not from hypercarbia. Hypercarbia and severe acidosis however, may impair cardiac performance, raise intracranial pressure and decrease efficacy of bronchodilators. The emphasis of desired gas exchange should be that of maintaining adequate oxygenation (O_2 saturation $\sim$95%) and allowing for permissive hypercapnia as long as pH is $\geq$ 7.3. Once corticosteroids and bronchodilator therapy decrease airway resistance as reflected in improved gas exchange, increased exhaled V_T and decreased autoPEEP, weaning could be started first by decreasing the PIP, decreasing mandatory breaths and gradually switching to pressure support ventilation.

Subsequent Course

Patient was managed on PCV mode under pharmacologic sedation and paralysis at a rate of 12/min, T_i of 1.2 s, T_e of 3.8 s, inflation pressure of 35 cm H_2O and PEEP of 4 cm H_2O. Clinical examination, ventilatory graphics, exhaled V_T, dynamic compliance and auto-PEEP were monitored. Permissive hypercapnia was allowed. Treatment with steroids and bronchodilators was continued. He also received

intravenous magnesium sulphate. Intravenous fluid boluses of 0.9% saline (10 ml/kg) were administered 3 times with improvement in his perfusion. Over the next 18 h, his dynamic compliance improved along with his gas exchange. Inflation pressure was decreased, pharmacologic paralysis was discontinued, and he was switched to pressure support ventilation while decreasing mandatory breaths. He was extubated 24 h after intubation.

Important Points

1. Diseases with increased resistance have prolonged time constants. Longer time is necessary for both delivering the V_T and alveolar emptying. Expiratory time constant is prolonged more than the inspiratory time constant. Mechanical ventilation should be performed at a relatively low mandatory rate and with higher inflation pressure or V_T.
2. EPP is displaced distally in intrathoracic airway obstruction resulting in widespread airway collapse and auto-PEEP. The consequences of auto-PEEP include decreased dynamic compliance and venous return. Application of extrinsic PEEP and longer T_e will help decrease auto-PEEP.
3. Sustained inflation pressure, either with PCV or PRVC results in better distribution of V_T compared to primarily volume controlled ventilation.
4. Monitoring of flow waveforms and auto-PEEP along with dynamic compliance is very helpful in determining the most effective and least harmful combination of mandatory breaths and inflation pressure (or V_T).
5. The goal of mechanical ventilation should be to maintain adequate oxygenation, reasonable pH and minimize complications.

13.4 Parenchymal Lung Disease

Case 4

A 16-month-old previously healthy girl presents to the emergency department with fever and progressively increasing respiratory difficulty over the past 12 h. Vital signs show a temperature of 39.5 °C, respiratory rate of 50/min, heart rate of 144/min, blood pressure of 70/36 mmHg and SpO_2 of 75% while breathing room air. Examination shows an anxious, irritable, toxic appearing child, with cool extremities and delayed capillary refill time. Appropriate fluid resuscitation and intravenous antibiotics were given. FiO_2 of 1.0 was administered via a high flow nasal cannula. There was no significant improvement in SpO_2. The decision to institute mechanical ventilatory support was made. The patient was pre-oxygenated with 100% O_2, given sedation and paralytic agents while providing bag and mask ventilation with a PEEP valve set at 10 cm H_2O.

After stabilization, endotracheal (ET) intubation was performed. Bag to ET tube manual ventilation was continued with 100% O_2 and PEEP of 10 cm H_2O, at a rate

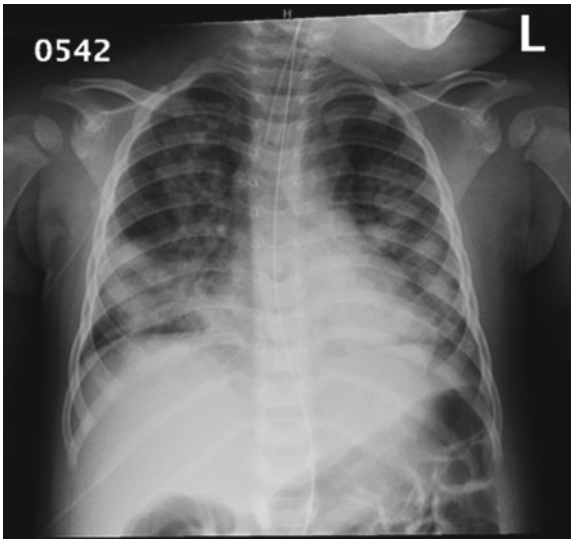

Fig. 13.3 A 16-month-old girl with fever and respiratory distress. Radiograph shows bilateral fluffy infiltrates in the lower lobes with extension in middle and upper lobes. Lung volume is decreased

of 30–40/min with an I:E ratio of approximately 1:1. SpO_2 improved to 96% and the patient was transported to the ICU for further management. Chest radiograph showed ET tube at mid-tracheal level, decreased lung volume, and bilateral coarse and dense opacities throughout the lung but more in lower lung fields (Fig. 13.3). Pressure-regulated volume control (PRVC) ventilation was instituted to deliver 7 ml/kg tidal volume at a PIP of 30 cm H_2O, ventilator rate of 30/min, with a T_i of 0.8 s, PEEP of 10 cm H_2O, and an FiO_2 of 1.0.

Clinical Analysis

This child has severe parenchymal pathology with primary involvement of the alveoli. Most likely cause is pneumonia of bacterial origin. She also exhibits signs and symptoms of septic shock which mandates appropriate antibiotics and fluid expansion along with inotropic support. The pathophysiologic consequences of the pulmonary involvement include reduction in FRC and decreased time constants for both inflation and deflation. Decrease in FRC will result mainly in hypoxemia since the pulmonary capillary blood will have less O_2 to pick up during exhalation. Critical opening pressure is increased for the alveoli. Reduction in alveolar pressure during exhalation carries the risk of alveolar collapse (de-recruitment) and alveolar-airway junction stress during inspiration (tidal recruitment), the main cause of volutrauma. Decreased lung compliance with relative sparing of airway resistance implies shortened time constants. The airway pressure and alveolar pressure take relatively shorter time (compared to normal) to equilibrate with each other. The end result is that both inflation and deflation will be completed relatively quickly. The pressure volume relationship with this type of mechanical abnormalities can be represented by an early phase where less volume change occurs for a given pressure ($\Delta V/\Delta P$), followed by a lower inflection point with an increasing $\Delta V/\Delta P$ with

another inflection point after which there is decreasing $\Delta V/\Delta P$. Thus, the dynamic compliance is greatest between the two inflection points. The alveolar pressure below the lower inflection point reflects de-recruitment (atelectrauma) and above the upper inflection point represents overdistension (volutrauma). Keeping PEEP below the lower inflection point carries the risk of alveolar de-recruitment and opening them again with an inspiratory pressure—a process sometimes referred to as "tidal recruitment"- exposing the delicate alveolar-terminal airway junctions to shear stress and rupture. PIP beyond the upper inflection point will subject the lungs to unnecessary volutrauma or barotrauma. The challenge to the clinician is to keep the tidal ventilation between these two points, by providing sufficient PEEP to keep alveoli recruited and limiting peak inflation pressure to avoid overdistension. The predominant gas exchange abnormality is hypoxemia. The safest strategies to manage hypoxemia are (a) an adequate PEEP and (b) increasing the inspiratory time (T_i). PEEP will help establish FRC and allow the pulmonary capillary blood to equilibrate with a higher PAO_2. The optimal PEEP can be inferred with improvement in PaO_2/FiO_2 and dynamic compliance. Both these measurements are helpful in keeping the tidal ventilation in between the two inflection points. Increasing Ti will allow the pulmonary capillary blood to equilibrate with the higher PAO_2 during inspiration while still enabling complete exhalation because of the short time constant. Weaning the FiO_2 to maintain sufficient SaO_2 ($\sim$95%) will reduce oxytrauma.

Subsequent Course

Patient was managed with PRVC to deliver V_T of 7 ml/kg, rate of 30/min, T_i of 0.8 s, PEEP of 10 cm H_2O and PIP limit of 35 cm H_2O. C_{DYN} was frequently monitored to make appropriate adjustments of PEEP. Pharmacologic sedation and paralysis were introduced to minimize asynchronous ventilation. Blood culture yielded methicillin sensitive *S. aureus*. The patient had a stormy clinical course requiring inotropic support and intravenous fluid expansion. Pulmonary infiltrates worsened with decreasing compliance and PaO_2/FiO_2 ratio, and greater need for oxygen and PEEP. High frequency oscillatory ventilation was employed with MAP of 26 cm H_2O, FiO_2 of 1.0, amplitude pressure of 30 cmH_2O and a frequency of 6 Hz. A steady improvement occurred with an increase in PaO_2/FiO_2 and lung aeration. She was transferred back to PRVC mode of ventilation when her MAP requirement was around 15 cm H_2O. Pharmacologic paralysis was discontinued. FiO_2 was weaned to keep $SpO_2 \sim 95\%$, mandatory rate was decreased to allow for pressure support ventilation. Patient was successfully extubated after a total of 10 days of mechanical ventilation.

Important Points

1. Time constant is decreased in diseases characterized by reduced compliance. This means that the proximal airway pressure and alveolar pressure equilibrate with each other in relatively a short period of time.

2. The major pathophysiologic consequence of alveolar/parenchymal diseases is decreased FRC and hypoxemia.
3. Alveolar recruitment by restoration of FRC with PEEP is a major therapeutic strategy.
4. Ventilation with low PIP/V_T at an appropriate FRC is a proven method of lung protection in pulmonary parenchymal disorders such as ARDS and pneumonia.
5. V_T should be delivered between the lower and upper inflection points. This can be determined by calculation of C_{DYN} at various levels of PEEP.
6. Increase in T_i can lead to improved oxygenation by increasing the time available to pulmonary capillary blood for gas exchange with higher PAO_2 during inspiration.
7. Patient disconnection with PEEP such as during suctioning carries the potential risk of alveolar de-recruitment and hypoxia. Such procedures should be performed while PEEP is being maintained.

13.5 Restrictive Chest Disease

Case 5

A 14-year-old boy with cerebral palsy and severe scoliosis is admitted with fever, tachycardia, hypotension, and poor peripheral perfusion. Clinical evaluation revealed a urinary tract infection with elevated inflammatory markers (white blood cell count, C-reactive protein, and procalcitonin) and combined respiratory and metabolic acidosis. An ABG revealed a pH of 7.06, $PaCO_2$ of 80 mmHg, PaO_2 of 80 mmHg, HCO_3 of 24 meq/L and a Base excess of −8 meq/L with a lactate level of 4 mmol/L. He also presented with a decreased mental status from baseline and respiratory distress with increased work of breathing.

Q. What would be the appropriate response at this time?

1. High-flow nasal cannula
2. Non-invasive ventilation with BiPAP
3. Intubation and invasive mechanical ventilation.

Answer: He definitely has acute on chronic respiratory failure with acidemia. While high flow nasal cannula might reduce his work of breathing and decrease his dead space, it is unlikely to correct his respiratory failure back to baseline. Noninvasive ventilation would not be recommended since he has decreased mental status from baseline. Therefore, the best response would be intubation and invasive mechanical ventilation.

The patient was intubated and placed on invasive mechanical ventilation. His chest x-ray showed the ET tube to be in mid-tracheal position, severe scoliosis, with small lung volumes (not different from prior films when he was well), the right lung being

considerably smaller than the left lung, with no infiltrates or effusion. His initial settings were a tidal volume of 8 mL/kg, a rate of 16/min, FiO_2 of 1.0, a PEEP of 5 cm H_2O, with an inspiratory time of 1 s on volume control ventilation. His peak inspiratory pressure was 40 cm H_2O with a plateau pressure of 35 cm H_2O. His ABG an hour after mechanical ventilation was pH of 7.20, $PaCO_2$ of 70 mmHg, PaO_2 of 350 mmHg, HCO_3 of 26 meq/L, and a base excess of −7 meq/L.

Clinical Analysis

The patient has urosepsis with acute on chronic respiratory failure with combined respiratory and metabolic acidosis. His severe scoliosis has produced a restrictive chest disease characterized by low lung volumes, decreased respiratory system compliance despite a normal looking lung on chest x-ray and chronic hypercarbia. There are many aspects of physiology to consider in optimizing mechanical ventilation in patients with restrictive chest disease. First, airway pressures are too high even with a tidal volume of 7 mL/kg on volume control ventilation. It would be preferable to limit the peak alveolar pressure by reducing the tidal volume. While the tidal volume can be reduced in volume control to limit the plateau pressure, the patient also has differential lung volumes due to severe scoliosis. The distribution of ventilation would be uneven and exacerbated by volume control ventilation. With a pressure-controlled time-cycled breath, the peak alveolar pressure would be limited to the same level in all alveoli, albeit with the lung segments having different time constants for inflation. Therefore, the preferred mode of ventilation in this patient would be pressure-controlled time-cycled ventilation. Secondly, by reducing the tidal volume, total minute ventilation would be decreased exacerbating the hypercarbia. Therefore, a much higher rate than 16 would be required to control the hypercarbia. Since he has chronic respiratory failure, his $PaCO_2$ does not have to be reduced to normal levels. Since he has no lung disease, it would not be necessary to increase his PEEP any further. His FiO_2 can be safely decreased to less than 0.5 and weaned from there as tolerated. When his sepsis is under control and he is being weaned, it might be prudent to extubate him to noninvasive mechanical ventilation.

Subsequent Course

He was extubated to noninvasive mechanical ventilation. Since this critical illness made him weaker, he needed noninvasive ventilatory assistance to maintain his gas exchange. He was weaned to intermittent BiPAP support and discharged on intermittent BiPAP support without any supplemental oxygen. About a month later, he was able to wean off the intermittent BiPAP.

Important Points

1. The choice of respiratory support must be tailored to the patient's condition. Invasive mechanical ventilation would be the choice for management for respiratory failure when there is altered mental status with inability to protect the airways from aspiration of secretions

2. Restrictive chest disease in this patient is due to several factors including scoliosis, ankylosis of costovertebral joints as well as sternocostal joints. With the result, the chest does not expand normally. Even with a lung protective strategy with a tidal volume of 6–7 mL/kg, the peak inspiratory and peak alveolar pressures reached in the lung may be too high. Therefore, these patients may need an even lower tidal volume. However, the ventilator rate has to be increased to maintain adequate minute ventilation.
3. With scoliosis, even though the lungs are relatively normal, lung expansion is asymmetric between the two lungs due to different time-constants between the two lungs. Volume-controlled ventilation, in this circumstance, results in uneven distribution of tidal volume and may result in atelectasis in the lung of the stiffer chest cavity while causing overexpansion of the compliant thorax. Therefore, the preferred mode of ventilation for this clinical situation is pressure-controlled time-cycled ventilation with an inspiratory time sufficiently long for adequate inflation.
4. In patients with chronic CO_2 retention, it is not necessary for ventilatory support to normalize $PaCO_2$. Permissive hypercapnia is the appropriate management strategy targeting a $PaCO_2$ closer to his baseline or higher, whichever is necessary to provide the most lung protection.
5. It might take longer for many of such patients to regain baseline respiratory function requiring a higher level of support after discharge from the ICU to adapt to the altered respiratory function. Home management with BiPAP may be necessary for some time before the previous respiratory status is established.

13.6 Respiratory Pump Disorder

Case 6

A 16-year-old male with no past medical history presented with a 1-day history of progressive lower limb weakness and paresthesia. He felt tired the night before when he went to bed. Morning of admission, he was able to walk but with support. There was no back pain, upper limb weakness, diplopia, dysphagia or shortness of breath or trauma to the spine. A week before his current visit, he had a fever and a cold which lasted about 3 days. Upon initial examination, he was alert, conscious and not in respiratory distress. His presenting vital signs were as follows: blood pressure of 120/68 mmHg, heart rate of 85 beats per minute (bpm), temperature of 37 °C, SpO_2 of 96% on room air. His lower limb examination revealed a muscle power of 1/5, absent tendon reflexes at the patellae and ankles and bilateral plantar flexion response to plantar reflex. He is now unable to walk even with support. Rest of the neurologic examination is unremarkable. No spinal tenderness is noted. His complete blood count, renal profile with electrolytes, liver function, cardiac enzymes, blood gas and random blood sugar tests are within normal limits. His

electrocardiogram is normal. His lumbar puncture shows 5 cells/mm^3, glucose of 75 mg/dL with a serum glucose of 110 mg/dL, protein of 220 mg/dL. Gram stain is negative. MRI of the spine shows surface thickening and contrast enhancement on the conus medullaris and the nerve roots of the cauda equina.

Differential diagnosis would include Guillain–Barre syndrome (GBS), transverse myelitis, or acute demyelinating myelitis. Since there is no sensory loss, transverse myelitis would be ruled out. Additionally, the typical MRI appearance in transverse myelitis is a central T2 hyperintense spinal cord lesion extending over more than two segments, involving more than two-thirds of the cross-sectional area of the cord. Acute demyelinating myelitis would also show lesions in the spinal cord as opposed to the nerve roots. Nerve conduction abnormalities in GBS would include slow or blocked nerve conduction, prolongation of distal latency and f-waves. His clinical course is most consistent with acute motor axonopathy, a variant of GBS.

Q. Where should this patient be admitted and what monitoring is required for this patient?

Answer: He has rapidly progressive weakness within a day. He is likely to develop worsening ascending paralysis. The concern would be the weakness affecting his respiratory muscles. Therefore, he needs to be admitted to the ICU where he can be monitored closely. His monitoring should include respiratory muscle strength assessment. There are 2 tests that are commonly employed to evaluate the respiratory muscle strength. One is the maximal inspiratory pressure that can be generated by the patient. The other is vital capacity. Maximal inspiratory pressure is measured by using the digital vacuum manometer attached to a unidirectional expiratory valve and a face mask or a mouth-piece. Patients will be able exhale through the valve, but inspiration will close the valve and generate a negative pressure. Care must be taken so as not to permit the negative pressure generated by the sucking action of the buccal muscles to influence the measurement. For this, an appropriate mouth piece is required to negate the action of the buccal muscles. Normal maximal inspiratory pressure is about −60 to −80 cm H_2O. Indication for intubation is a maximal inspiratory pressure of −20 cm H_2O or higher (less negative).

Vital capacity measurement also requires a maximal respiratory effort from the patient. Volumes are measured using a spirometer that can be attached to a mouth piece or a face-mask. Patients are encouraged to make a maximal inspiration followed by a maximal exhalation. The exhaled volume from maximal inspiration to exhalation is the vital capacity. Normal vital capacity is 60–80 mL/kg. A vital capacity of 15 mL/kg is an indication for mechanical ventilation.

On admission, the patient's maximal inspiratory pressure was −50 cm H_2O and his vital capacity was 40 mL/kg. Nerve conduction studies confirmed the diagnosis of GBS. He was treated with IVIG. Six hours later, he was complaining of air hunger and dyspnea. His maximal inspiratory pressure was −15 cm H_2O and his vital capacity was 15 mL/kg. His SpO_2 was 95% in room air and a blood gas showed a $PaCO_2$ of 50 mmHg. A decision was made to provide mechanical ventilation.

Q. Would non-invasive mechanical ventilation be appropriate for this patient?

Answer: This patient has rapidly progressive respiratory compromise. Therefore, noninvasive ventilation would not be appropriate for this patient.

Subsequent Course

The patient was intubated and placed on invasive mechanical ventilation. A volume controlled (V_T 10 mL/Kg) SIMV mode at rate of 16/min was used with a pressure support of 10 cm H_2O. He was treated with IVIG for 5 days with improvement in his muscle weakness. While he was being ventilated his maximal inspiratory pressure and vital capacity were measured daily.

Q. What would appropriate levels of maximal inspiratory pressure and vital capacity to consider weaning towards extubation?

Answer: His maximal inspiratory pressure should be at least −20 cm H_2O and vital capacity should be at least 20–30 mL/kg before weaning towards extubation is commenced. If his maximal inspiratory pressure and vital capacity are closer to normal, then he could be weaned and extubated to complete spontaneous breathing without any positive pressure assistance. If his measurements are low but above the threshold stated above, the patient could be extubated to noninvasive mechanical ventilation provided that wakefulness and airway protective reflexes are adequate.

Important Points

1. Rapidly progressive muscle weakness is an indication for ICU care even if the patient at the time of admission does not require any positive pressure support. The more frequent and intensive monitoring available in the ICU will allow the appropriate respiratory support to be provided as soon as necessary for the patient.
2. In a potentially progressive neuropathy, inspiratory muscle strength and capacity needs to be monitored closely and frequently. The decision to intubate would be made more by the index of inspiratory muscle strength and capacity rather than blood gases. Blood gases may be normal before the inspiratory muscle strength reaches the threshold for intubation.
3. Rapidly progressive muscle weakness especially with the ascending involvement of the intercostal muscles is a contraindication for noninvasive ventilation. Noninvasive ventilation, under these circumstances, may delay intubation and can result in an emergent situation with its attendant complications and morbidity.
4. Once the patient is on invasive ventilation, the clinical progress is monitored by serial measurement of inspiratory muscle strength and capacity. When the inspiratory muscle strength has improved sufficiently, the patient should be considered ready to be weaned off mechanical ventilation.

13.7 Abdominal Distension

Case 7

A one-year-old with biliary atresia, s/p Kasai procedure with portal hypertension and ascites is admitted with viral pneumonia. Chest x-ray shows bilateral diffuse patchy infiltrates mainly in the lower lobes. He is intubated and placed on mechanical ventilation. His initial ventilator settings are a tidal volume of 6 mL/kg, PIP of 30 cm H_2O, PEEP of 6 cm H_2O, FiO_2 1.0, rate of 25/min, and an inspiratory time of 0.8 s in pressure-controlled time-cycled ventilation. He is sedated and paralyzed. His first ABG shows: pH 7.36, $PaCO_2$ 50 torr, PaO_2 of 70 torr, HCO_3 25 meq/L, base excess −1 meq/L. His SpO_2 is 95%. His abdomen is distended and tense with an umbilical hernia and a fluid thrill. His chest x-ray shows the ET tube to be in the mid-tracheal position, with bilateral infiltrates (lower lobes > upper lobes), elevated diaphragm, and 7 rib expansion.

Case Discussion

From a respiratory point of view, he has severe ARDS (PaO_2/FiO_2 ratio < 100) with adequate ventilation (mildly hypercarbic). He has decreased lung volume, and his oxygenation is adequate.

Q. What should the next steps be?

1. Repeat a blood gas in an hour before making any change in ventilator settings
2. Accept current settings and gas exchange
3. Start inhaled nitric oxide
4. Increase PEEP to 8 cm H_2O.

Answer: Given the fact that his lung volume is reduced and he has severe oxygenation failure, the appropriate next step is to recruit the lungs and maintain the recruitment. That can be reliably done with an increase in PEEP. Repeating a blood gas in an hour is only postponing the necessary change in ventilator settings since the gas exchange is unlikely to change in an hour. Accepting the current settings in the setting of de-recruited lungs is not an optimal solution. Inhaled nitric oxide is not appropriate as there is no evidence of pulmonary hypertension.

Case Progression

PEEP was increased to 8 cm H_2O. A repeat ABG showed a pH of 7.35, $PaCO_2$ of 52 torr, PaO_2 of 72 torr, HCO_3 of 24 meq/L and a base excess of −2 meq/L. SpO_2 was 95%. Why did this patient not respond to an increase in PEEP? The reasons are either the patient requires a higher level of PEEP or that his lungs are not recruitable. This patient has abdominal distension as well as an increase in intraabdominal pressure. Increased abdominal pressure restricts the amount of lung inflation in the lower lobes close to the diaphragm. When the airway pressure at the bases is less than the intrabdominal pressure, the lung segments exposed to the intrabdominal pressure will collapse. In order to open these lung segments, one must apply a

pressure that is equal to or preferably slightly higher than the intrabdominal pressure. There are two ways one can find the appropriate level of PEEP for this patient. One is to measure the intrabdominal pressure and to set the PEEP just a couple of centimeters above the intraabdominal pressure. Intrabdominal pressure can be measured either by measuring the bladder pressure through a urinary catheter or intragastric pressure through a nasogastric tube. The second method is to do a bedside PEEP titration. With this method, PEEP is increased in steps of 2 cm H_2O to observe the delivered and exhaled tidal volumes with a change in PEEP. In a recruitable lung, as PEEP is increased, tidal volume will also increase in pressure-controlled ventilation due to an improvement in lung compliance. The best PEEP would be the level at which the maximum increase in tidal volume was observed. If the lung is not recruitable, then there would either no change in tidal volume or a decrease in tidal volume if the lungs are hyperinflated. In the case of increased abdominal pressure, as PEEP is increased, one may see an abrupt increase in tidal volume when the PEEP level is above the intrabdominal pressure. This patient had both maneuvers performed. His bladder pressure was 12 cm H_2O. During the PEEP trial, with a change in PEEP from 6 to 8 to 10 to 12 cm H_2O, no change in tidal volume was observed which was set at 6 mL/kg. When PEEP was increased from 12 to 14 cm H_2O, tidal volume increased to 9 mL/kg, and further increased to 10 mL/kg when PEEP was increased to 16 cm H_2O but at this level of PEEP, his blood pressure decreased and heart rate increased. His PEEP was set at 14 cm H_2O. His PIP could be reduced to 30 cm H_2O to maintain a tidal volume of 6 mL/kg. His repeat ABG showed a pH of 7.36, $PaCO_2$ of 50 torr, PaO_2 of 245 torr, HCO_3 of 24 meq/L and a BE of −2 meq/L. This case illustrates the concept that the transpulmonary pressure, the alveolar pressure minus the surrounding pressure should be greater than zero for the alveoli to remain open. In patients, with increased abdominal pressure, increasing PEEP can have negative hemodynamic effects. Therefore, the optimal level of PEEP would be the one that results in improvement in lung mechanics and gas exchange while balancing the negative effects on the circulation. Increases in PEEP can however, adversely affect venous return and cardiac output. To counteract this, abdominal pressure should be reduced by paracentesis. Paracentesis can be associated with hypotension due to hypovolemia and may require an appropriate volume infusion. Decreasing abdominal pressure can also increase transpulmonary pressure by decreasing the forces opposing alveolar pressure.

Important Points

1. In order for an alveolus to remain open, the transpulmonary pressure (alveolar pressure minus the pleural pressure) must be positive. If the transpulmonary pressure is less than zero, it will result in atelectasis of the involved alveoli and contribute to intrapulmonary shunting.
2. With abdominal distension, intrabdominal pressure increases. This means that the alveoli exposed to the abdominal pressure near the diaphragm need a higher pressure to overcome the abdominal pressure. During inspiration, the inspiratory pressure may be higher than the abdominal pressure and open the alveoli. But,

during exhalation, if the PEEP level is below the intraabdominal pressure, the alveoli will collapse and contribute to venous admixture.

3. Repeated opening and closing of alveoli will contribute to atelectotrauma. To avoid this, alveoli need to be maintained open both during inspiration and exhalation. Setting the PEEP as described above, so that the transpulmonary pressure is positive throughout the respiratory cycle, is an important component of ventilator management.
4. Moderate to severe intraabdominal pressure increase, especially due to fluid accumulation, may require paracentesis to drain the fluid and decrease the intraabdominal pressure.
5. It is important to remember that both paracentesis and PEEP titration are strategies to increase the transpulmonary pressure and may need to be combined in many patients.
6. Removal of fluid from the abdomen, especially when it is under high pressure, may result in hypotension due to hypovolemia requiring the restoration of circulating blood volume. It would be prudent to administer the fluid before paracentesis is performed to mitigate the decrease in blood pressure.