

# Chest wall disorders & the lung:

- A group of conditions affecting the Inspiratory pump and potentially leading to respiratory failure.
- Disorders may affect all mechanical components of the Inspiratory pump.
  - Respiratory muscles
  - Bony rib cage
  - The spine and its articulations.
  - Soft tissue compromising the abdomen
- Lead to a mechanical disadvantage to the Inspiratory pump and an increased work of breathing.
- Characterized usually by a restrictive defect.
- Share the potential of long term hypercapnic respiratory failure

## Etiology of chest wall disorders:

### 1. Spine & articulations:

Kyphoscoliosis

Thoracoplasty

### 2. Rib cage:

flail chest( esp with pulmonary contusion.)

Pectus excavatum/

Pectus carinatum

Ankylosing spondylitis.

### 3. Respiratory muscle involvement:

Central

Peripheral: nerve/ neuromuscular junction/ muscle.

### 4. Soft tissue: obesity.

Kyphosis may exist as an isolated entity.

Osteoporosis.  
post-radiation.

Estimated prevalence: mild deformity 1 in 1000  
severe deformity 1 in 10000 in U.S

Scoliosis invariably associated with Kyphosis.

### A. Congenital

Isolated

Associated syndromes: NF/ Marfan's /EDS.

### B. Paralytic/ Secondary

Neuromuscular: poliomyelitis/ M. dystrophy /CP/CMT syndromes.

connective tissue related: EDS/ Morquio's.

vertebral disease: osteoporosis/ osteomalacia/ spina bifida.

compensatory: post Thoracoplasty.

### C. Idiopathic.

## Idiopathic Kyphoscoliosis

- Often begins in early adolescence a female predominance (4 to 1)
- May account for as many as 80% of cases of Kyphoscoliosis
- The Cobb angle is associated with the degree of restriction. The relationship is however more complex.

Other factors involved are:

1. Number Of Vertebrae
2. Location of curve
3. Patient's age.
4. Presence of Kyphosis
5. Degree of rotation

## **Congenital Kyphoscoliosis:**

Isolated or Syndromic/may be genetic

present at birth

Malformations of vertebrae( during ontogeny)

loss of VC 15% greater than in idiopathic scoliosis.

## **Paralytic Kyphoscoliosis:**

loss of muscular tone primary

secondary unequal resting tone & remodeling.

should be ruled before other 2 categories considered.

association between scoliosis & restriction not strong.

reduction of VC primarily assoc with weakness.

early onset of scoliosis.

faster progression.

# Pathophysiology of Kyphoscoliosis

Maximum restriction among chest wall disorders.

## Pulmonary function testing:

↓ TLC

↓ VC

N/ ↑ RV

N FEV1/FVC

↓ FEV1

↓ RV/TLC.

Mimics obstructive  
disease.

FEV1 reduced proportional to the restriction.

Specific airway conductance is actually increased( radial traction because of ↑ recoil.)

FEV1 is grossly reduced in a scoliotic patient?

Think of

- 1.torsion of central airways
- 2.impingement of mediastinal structure( eg aorta) on the airways.
- 3.smoking related obstructive airways disease.

$DL_{CO}$

both steady-state and single-breath measurements are often reduced in proportion to the reduction in TLC.

$DL_{CO}$  in some patients may be reduced disproportionately

- 1.failure of alveolar development or
- 2.anatomic changes in the pulmonary circulation.

(usually elderly patients with cor pulmonale and Cobb angles > 120 degrees)

## Physiology Of Kyphoscoliosis :

Respiratory system compliance is reduced in Kyphoscoliosis.

1. Primarily due to a reduction in chest wall compliance and to a lesser degree, reductions in lung compliance.

<50 degrees :minimal effect in decreasing respiratory system compliance

>100 degrees decrease the respiratory system compliance grossly.

2. FRC is at a lower lung volume. This further reduces respiratory system compliance by shifting tidal breathing to a flatter portion of the volume-pressure curve.

3. In children with even severe Kyphoscoliosis, chest wall compliance is normal, and impairment in ventilatory function may be due to mechanical inefficiency of the unequal hemi thoraces

4. The reduction in lung compliance due to micro atelectasis . Not due to intrinsic lung disease.

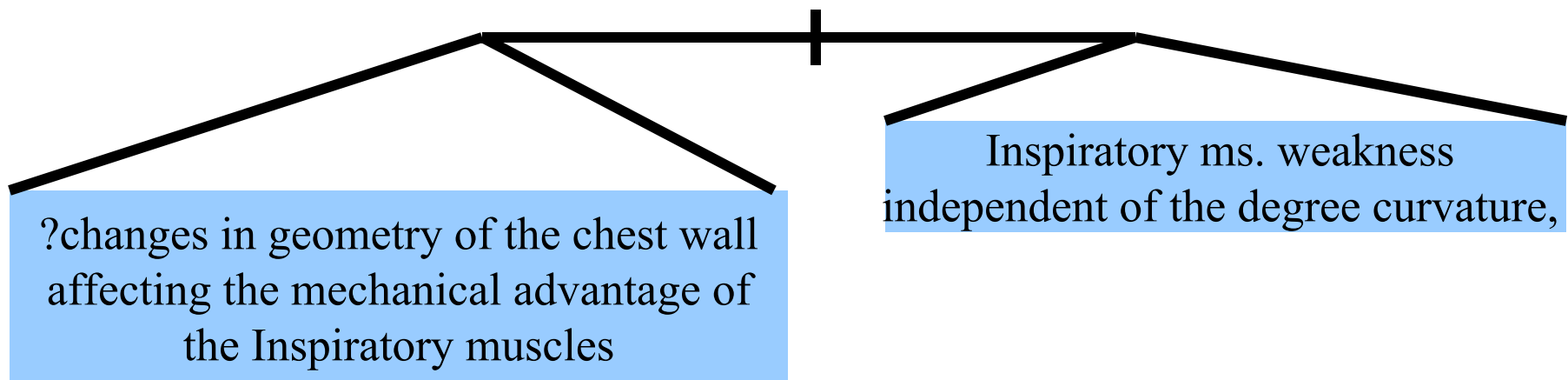


- Reduction in respiratory system compliance ↑ the elastic load placed on the respiratory muscles increasing WOB  
twice the normal value to inflate the lung and  
five times the normal value to inflate the chest wall.
- WOB is positively related to the oxygen cost of breathing. Increased three to five times that seen in healthy subjects.
- Such increase in the oxygen cost of breathing may place these patients at risk for respiratory muscle fatigue.
- Intercurrent illnesses may pose additional elastic loads, further increasing the work and oxygen cost of breathing

## **Respiratory Muscle Strength**

In paralytic Kyphoscoliosis, Inspiratory muscle weakness contributes to the restrictive process

- In patients with idiopathic Kyphoscoliosis, the prevalence of respiratory muscle weakness and its role in the development of respiratory failure is unknown.
- $PI_{max}$  and  $PE_{max}$  is normal in young patients with idiopathic scoliosis with Cobb angles of less than 50 degrees.
- With Cobb angles  $>50$  degrees, both  $PI_{max}$  and  $PE_{max}$  are mildly decreased to 70% and 80% of control values, respectively.
- In older patients with somewhat more scoliosis,  $PI_{max}$  is about 50% of predicted in eucapnic patients and 25% of predicted in hypercapnic patients.



## Control and Pattern of Breathing

Compensation for the reduced compliance & the increased elastic load on the Inspiratory muscles

1. recruit the Inspiratory muscles of the rib cage and
2. adopt a rapid shallow breathing pattern consisting of a low tidal volume and shortened Inspiratory time.

Advantages of breathing with low tidal volumes:

1. Reduction in the work per breath.
2. Reduces the ratio of the pressure needed to inhale ( $P_{\text{breath}}$ ) to  $P_{\text{Imax}}$ , thereby lowering the risk of developing Inspiratory muscle fatigue and reducing dyspnea.

Disadvantages of adopting a low tidal volume breathing

1. the potential for developing microatelectasis
2. increased dead space ventilation.

- Alternatively or concurrent compensation includes increasing central neural drive to the muscles.( as measured by mouth occlusion pressure P0.1 ).
- The ventilatory response to carbon dioxide, however, may be normal or reduced.
- If the response is reduced, the magnitude of the reduction is proportional to the degree of mechanical limitation imposed by the chest wall .
- External resistive loading than inherent problem with respiratory control.
- The drive to breathe in these patients is normal, but compensatory increases in minute ventilation are limited by mechanical factors.
- The effects of aging and its influence on central ventilatory control unknown.

## Gas Exchange

Hypoxemia may be due to

- 1.ventilation-perfusion mismatch (under ventilation of lung regions)
- 2.underlying atelectasis, or
- 3.alveolar hypoventilation.

Independent of the side of the hemi thorax relative to the scoliosis esp. with Cobb angles  $>65$  degrees

AaDO<sub>2</sub> less than that seen in normocapnic patients with COPD (15 vs. 40 mm Hg).

When hypoventilation supervenes, hypoxemia worsens and hypercapnia develops.

Hypercapnia initially appears during sleep and with exercise. Eventually, with further disease progression, it occurs at rest.

Causes of Pulmonary Hypertension.

- 1.Persistent hypoxemia leads to pulmonary vasoconstriction , RVH and cor pulmonale.
2. Proliferation of pulmonary artery smooth muscle
- 3.compression of the pulmonary vascular bed by a deflated lung.

## Exercise Capacity

- Exercise capacity significantly impaired in patients with severe Kyphoscoliosis.
- Maximum oxygen consumption reduced to about 60% to 80% of predicted
- However, the breathing pattern response to exercise with Cobb angle between 25 and 70 degrees is similar to that seen in normal subjects.
- The ratio of tidal volume to vital capacity (VT/VC) is roughly 0.5
- ratio of  $\dot{V}E_{max}$  /MVV can reach 70%.
- Supplemental oxygen & Corrective surgery during exercise may increase oxygen saturation and decrease dyspnea but it does not affect walking distance.
- Deconditioning and lack of regular aerobic exercise may also contribute to poor exercise performance in adolescents and adults with moderate scoliosis.

## Sleep-Disordered Breathing

The most common abnormality during sleep is hypoventilation.

1. Hypotonia of the intercostal and accessory muscles, particularly during rapid-eye-movement (REM) sleep
2. Decreased neural drive to the diaphragm during REM sleep
3. Decreased chest wall compliance. May not correlate with the degree of thoracic deformity or the severity of the restrictive process.
4. Hypoxemia-related autonomic dysfunction manifested by pronounced variability of heart rate.
5. Coexistent obstructive sleep apnea. Distortion of the upper airway anatomy may predispose these individuals to intermittent airway obstruction during REM-induced hypotonia of the pharyngeal muscles.

Recurrent hypercarbia and hypoxemia during sleep portends the daytime development of hypercarbia, cor pulmonale, and respiratory muscle dysfunction and cardiorespiratory failure.

Potentially treatable or reversible cause of respiratory failure.

## Clinical features of Kyphoscoliosis

Five major presentations in the natural history of untreated adolescent idiopathic scoliosis in adults are:

- (1) back pain,
- (2) psychosocial effects,
- (3) curve progression
- (4) Cardiopulmonary complications and
- (5) Mortality.

Congenital and secondary scoliosis often noted by the physician during screening.

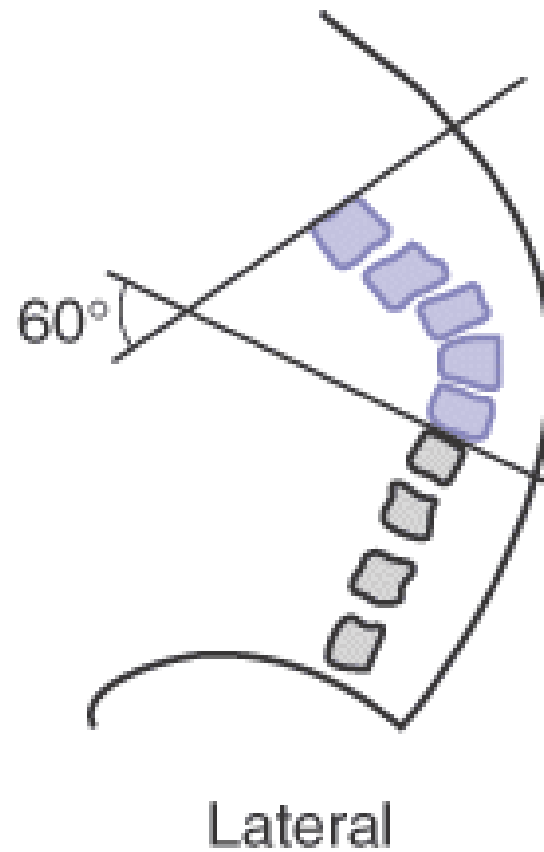
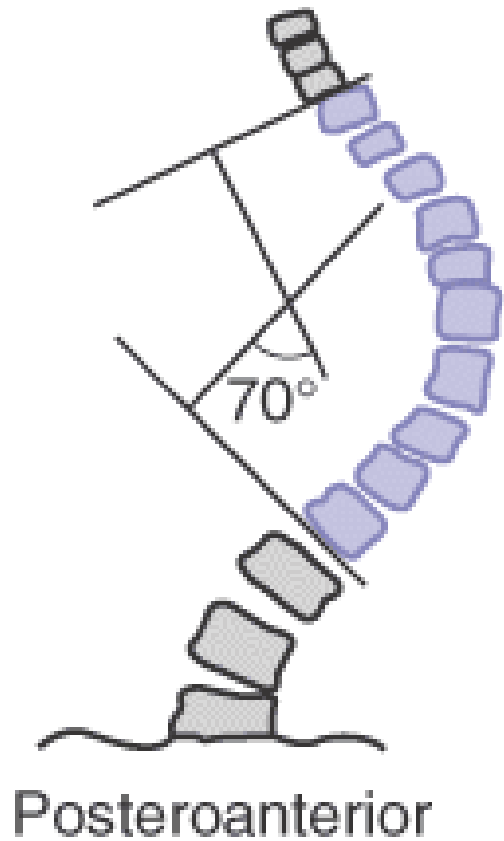
Congenital Kyphoscoliosis may be associated with neuro-deficit due to rapid progression. Secondary Kyphoscoliosis recognized as part of overall diagnosis.



## Cobb's angle: Measurement.

Simple investigation to quantify the degree of Kyphoscoliosis.

Requires only AP & Lat X-rays.



## Importance of the Cobb's angle:

- Angle formed by the intersection of two lines, each of which is parallel to the top and bottom vertebra of the scoliotic or kyphotic curves
- The magnitude of the Cobb angle has been used to predict development of respiratory failure and need for intervention.
- A Cobb angle  $>100$  degrees considered a severe deformity and more likely to be associated with respiratory failure.
- In idiopathic Kyphoscoliosis, the Cobb angle is associated with the degree of restriction
- Patients with an angle  $>60$  degrees are more likely to exhibit a restrictive defect. Those individuals with angles in the 90- to 100-degree range have severe restriction.
- In secondary Kyphoscoliosis reductions in VC correlate closely with muscle weakness than with the degree of spinal curvature.
- In patients with congenital scoliosis, for any given Cobb angle, the loss in VC is 15% greater than in patients with idiopathic scoliosis.

## 2. Progression of the thoracic deformity

Progression of the thoracic deformity depends on

1. skeletal immaturity and
2. the degree of deformity at the time of diagnosis.

After skeletal maturity, thoracic deformities

- <30 degrees are unlikely to progress,
- 30 to 50 degrees increase slowly by an 10 to 15 degrees.
- >50 degrees increase steadily at a rate of about 1 degree annually.

Mild to moderate idiopathic kyphoscoliosis & severe idiopathic kyphoscoliosis  
<35 years of age are similar to the general population

- with regard to symptoms,
- loss of lung volume with aging,
- life expectancy.

## Summary of the Factors Related to Progression of Adolescent Idiopathic Scoliosis:

- Girls > boys
- Premenarchal
- Risser sign of 0
- Double curves > single curves
- Thoracic curves > lumbar curves
- More severe curves

### 3. Ventilatory failure in Kyphoscoliosis

- Middle-aged patients with  $>100$  degrees  
tend to develop dyspnea and decreased exercise tolerance,  
have repeated acute respiratory infections,  
at risk for respiratory failure
- The onset of respiratory failure is insidious and incidence variable.
- Kyphoscoliosis accounts for less than 5% of chronic ventilatory failure in adults.
- After Cor pulmonale develops, the prognosis is poor; without treatment, death generally occurs within a year.
- Pregnancy usually poses no added risk for respiratory complications.
- With severe degrees of kyphoscoliosis and reductions in VC to less than 1 liter the risk for respiratory complications during pregnancy may be increased

Prime factor that has been historical related to progression to respiratory failure has been the degree of kyphoscoliosis.

This notion has been challenged by cases of survival into 70's with curves >100 without any specific therapy.

#### Other Factors pre-disposing to Ventilatory failure in Kyphoscoliosis:

spinal deformity >100 degrees

underlying neuromuscular disease

Inspiratory muscle weakness

sleep disordered breathing

airway compression

co-existent pulmonary disease.

## Medical management: General Measures

Both preventive and supportive measures.

1. Immunization,
  2. adequate hydration,
  3. prompt care of respiratory infections,
  4. avoidance of sedatives, and
  5. carefully monitored supplemental oxygen.
  6. Abstaining from smoking and
  7. maintaining body weight within desirable levels
  8. Physical training is encouraged to improve exercise capacity in sedentary patients.
- Chronic respiratory failure treated with
    - chest physiotherapy,
    - bronchodilators,
    - oxygen therapy, and
    - diuretics if needed.
    - Non-invasive ventilation.

## Noninvasive Ventilation for Kyphoscoliosis

### Q1. Who to give Long-Term NIV for Kyphoscoliosis?

- A. Prophylactic therapy in high-risk patient
- B. As definite therapy for respiratory failure.

Definite therapy for respiratory failure: Indications.

AJRCC 163;2001:540

Symptoms (e.g., fatigue, morning headaches, dyspnea)

or

signs of cor pulmonale

and one of the following:

1. Daytime arterial PCO<sub>2</sub>  $\geq 45$  mm Hg
2. Nocturnal oxygen saturation  $\leq 88\%$  for five consecutive minutes
3. Progressive neuromuscular disease with P<sub>I</sub>max  $< 60$  cm H<sub>2</sub>O or FVC  $< 50\%$  of predicted



- Continued debate about whether nocturnal ventilation is of any benefit to asymptomatic patients
  1. who either belong to a high-risk
  2. have abnormal diurnal blood gases.
- Currently no data for any firm indication in a prophylactic role.

## Q2.How to best deliver NIV?

Nocturnal negative-pressure ventilation has been administered with devices like cuirass and body-wrap ventilators

Generally preferred to tank ventilators.

The benefits of noninvasive nocturnal ventilation in patients with kyphoscoliosis appear to be the same whether a negative-pressure or positive-pressure ventilatory mode is used

## Pros of using positive-pressure devices

1. Better acceptance
2. More experience
3. Portable
4. Occupies less space
5. Not need of customization
6. Can treat both Kyphoscoliosis & associated Upper airway dysfunction at once.
7. Volume controlled & can be safely used in sicker patients.

## Cons of using positive -pressure devices

1. Usual problems of claustrophobia
2. Facial deformities associated.

Because of the rapid, shallow breathing pattern adopted by these patients, the ventilator triggering system should have a short response time to minimize patient-ventilator asynchrony.

## Cons of using negative-pressure devices

Usually delivered as pressure controlled ventilation.

- Negative-pressure ventilation may lead to patient-ventilator asynchrony.
- induction of upper airway obstruction during sleep
- the need to custom-fit a cuirass to the deformed chest wall, and
- the associated patient discomfort of having to lie supine throughout the night

## Pros of using negative-pressure devices

Historical often.

Institutional preference

?? Pressure cycled

1. fewer data for negative-pressure ventilation
2. include patient comfort related to flow-triggering systems,
3. ability to compensate for leaks, and
4. ability to adjust the time to peak inspiratory positive airway pressure

??Volume cycled devices

All advantages of the positive pressure ventilators.

## Q3. Are there any Benefits of Noninvasive Nocturnal Ventilation in Chest Wall Diseases??

### **Gas Exchange**

Increased PO<sub>2</sub>

Decreased PCO<sub>2</sub>

### **Hemodynamics**

Decreased pulmonary artery pressure

Increased right ventricular function

### **Mechanics**

Reduced work of breathing

Increased P<sub>I</sub>max

### **Sleep Hygiene**

Normalized sleep patterns

Fewer apneic episodes

### **Outcomes**

Fewer hospitalizations

Improved quality of life,  
relief of dyspnea

1. A 10-year analysis of ANTADIR Observatory  
1996;109;741-749 Chest
2. Gonzalez, Gloria Ferris et al  
2003;124;857-862 Chest
3. Annane D, Chevret S, Chevret S, Raphaël JC  
The Cochrane Library 2004, Issue 3

- Improvements in sleep architecture and normalization of blood gases may occur within the first few days of treatment,
- Improvements in pulmonary hemodynamics may take longer to evolve.
- Small improvements in vital capacity, respiratory muscle strength, and endurance have also been recorded. Dyspnea on exertion, daytime somnolence, and early morning headaches can all be alleviated.
- Long-term ventilatory support in patients with kyphoscoliosis and respiratory failure significantly reduces the number of days spent in hospital
- RCT's demonstrating survival benefit in patients with kyphoscoliosis and respiratory failure treated with NIV lacking.
- With historical controls, survival benefit with kyphoscoliosis and respiratory failure receiving noninvasive ventilation seen.
- Survival was 90% after 1 year and 80% after 5 years

# Thoracoplasty & the lung.

Entails removal of a number of ribs( upto 7-8), along with their intercostal muscles.

Collapses the underlying lung( and associated cavities.)

Was a common surgical procedure for tuberculous empyema & cavitory TB prior to the advent of systemic chemotherapy in the 1940-50's.

ARTIFICIAL PNEUMOTHORAX

PHRENIC N. DIVISION.

PNEUMOPERITINEUM

THORACOPLASTY

EXTRAPLEURAL PNEUMOLYSIS/ EXTRAPLEURAL PNEUMOLYSIS  
AND PLOMBAGE

Presently used in few instances (n=37):

Ann Th Surg 93:56: 903.

1. Persistent tubercular BPF.
2. Post-pneumonectomy empyema
3. Sputum positive Multi-drug resistant tuberculosis resistant to all drugs.
4. Atypical mycobacteria resistant to chemotherapy.

Currently this procedure has a mortality of <10% peri-op.

### **Current Indian experience in Thoracoplasty.**

*Indian J Chest Dis Allied Set 1999; 41 : 83-88 Departments of Thoracic Surgery and Anaesthesia  
L.R.S. Institute of TB and Allied Diseases, New Delhi*

- July 1992 and June 1997 , n=139 patients.
- tubercular empyema (84 patients),
- pyogenic empyema (33 patients),
- post-operative empyema with BPF(8 patients),
- drug resistant pulmonary tuberculosis (2 patients) and
- recurrent hemoptysis (2 patients).
- Successful outcome in the form of control of sepsis, closure of BPF, sputum conversion and control of hemoptysis was achieved in most cases. There were four deaths in the entire series.



## Basis of derangements in Thoracoplasty:

1. Reduced respiratory system compliance
2. Fibro thorax
3. Pulmonary fibrosis due to underlying granulomatous disease
4. Previous lung resection
5. Phrenic nerve injury
6. Progressive scoliosis
7. Inspiratory muscle weakness

- **Thoracoplasty** is second only to Kyphoscoliosis in producing a severe restrictive pattern
- VC and TLC may be reduced to 40% and 70% of predicted, respectively.
- RV and FRC are reduced to the range of 70% and 90% of predicted, respectively
- Progressive scoliosis and the effects of aging on respiratory system compliance worsen the restrictive process. The degree of restriction, however, does not correlate with the extent of Thoracoplasty
- Coincident airflow obstruction is common. FEV1 and the FEV1 /FVC ratio are reduced, and the RV/TLC ratio is increased
  - Airway obstruction may be due to
  - chronic bronchitis from cigarette smoking
  - previous tuberculous bronchitis or
  - bronchiectasis.
- Airway obstruction becomes more prevalent with aging and is associated with carbon dioxide retention, hypoxemia, and complaints of dyspnea.

- Of 171 patients who were operated on between 1951 and 1953, over one third had died by 1987.

**Philips, Kinnear. Thorax 42;1987:445.**

- Risk factors associated with developing respiratory failure include
  - preoperative tuberculous cavitory disease,
  - advanced age at the time of operation,
  - a preoperative contra lateral surgically induced pneumothorax, and
  - male gender.
  - Thoracoplasty than in those who underwent pneumonectomy alone.
- The maximal work rate and oxygen consumption are reduced, and there is a ventilatory limit to exercise (correlated with reductions in FEV1)
- The capacity to exercise, as measured by the 6-minute walk test, is roughly equal to that of individuals with COPD of similar age and comparable FEV1 During exercise, the diffusion capacity fails to increase appropriately, indicating an irreversibly restricted pulmonary vascular bed.
- Nocturnal hypoxemia is common in patients with hypercapnia or borderline low arterial PO<sub>2</sub> during daytime
- Cor pulmonale may develop as a result of hypoxemia and resultant pulmonary hypertension

# TREATMENT

- Treatment is supportive
  - domiciliary oxygen,
  - antibiotics when appropriate, and
  - noninvasive nocturnal ventilation.
  - In addition, any obstructive component should be vigorously treated
- Management of chronic respiratory failure often entails the use of positive-pressure or negative-pressure noninvasive ventilation.
- Non-invasive ventilation reduces the work of breathing, alleviates respiratory muscle fatigue, relieves dyspnea, lessens sleep-induced desaturation, and improves overall prognosis
- Improvements in VC, maximal inspiratory pressure, maximal expiratory pressure, and arterial blood gases occur soon after initiation of noninvasive ventilation and can be maintained over the long term.
- Even with nocturnal ventilatory assistance, however, mortality is still high, with a 55% survival rate 7 years after initiating treatment

## 3. Pectus & the chest:

### A. Pectus excavatum

- Pectus excavatum is characterized by excessive depression of the sternum. may be
  - minimal or extreme,
  - diffuse or local,
  - symmetrical or asymmetrical.
- Most common chest wall deformity seen by pediatricians and primary care providers affecting between 0.5% and 2.0% of the population
- Occurs in approximately 1 in every 1000 children.
- Boys are affected about three times more often than girls.
- Apparent at birth in over 80% of individuals The natural history is variable; usually progresses, especially during the teenage years.

- The etiology is unknown. A defect in the connective tissue surrounding the sternum has been implicated
- Frequent association with connective tissue disorders such as Marfan's syndrome.
- A genetic predisposition suggested by one large series in which a family history of Pectus deformity was present in 41% of the members. In most series, family history is not prominent.
- Associations with Pectus excavatum:
  - Scoliosis 15%,
  - congenital heart disease 4%,
  - functional heart murmurs 31%

## B. Pectus carinatum

- Pectus carinatum is a disorder in which the sternum is protuberant.
- Is less common than Pectus excavatum.
- It is of unknown etiology and may be associated with congenital heart disease, severe childhood asthma and rickets

- The most frequent complaints are cosmetic in nature and arise in patients between the ages of 15 and 20 years.
- Psychological problems related to the deformity occur in as many as 85% of patients.
- Dyspnea on exertion also occurs in 30% to 70% of patients. Other symptoms include chest pain, palpitations, and frequent respiratory infections
- Although extremely rare, respiratory failure can occur in adults with severe Pectus excavatum

### Objective assessment of Pectus disorders:

- A bedside determination of historical interest is the **hollow** index.
- This is the ratio between the amount of water contained in the chest depression and the body surface area. An index of greater than 50 mL/m<sup>2</sup> indicates the presence of pectus deformity

- The degree of chest wall deformity can be assessed clinically or radiographically. A **greater than 3 cm** distance between the surface of the anterior chest wall and the deepest sternal depression is considered significant
- A **minimal distance of <10 cm** between the posterior border of the sternum and the anterior border of the thoracic vertebra on the lateral chest radiograph is considered significant for Pectus. This measure, however, does not account for differences in body size
- To address this question, the ratio of the AP to transverse diameters of the rib cage (as seen on the poster anterior and lateral chest radiograph) is calculated. **A ratio of less than 0.4 indicates the presence of Pectus, and a ratio less than 0.3 indicates a severe deformity.**
- Anatomic definition can be obtained with CT scans. Measurements of transverse and AP chest wall diameters are taken at the level of the deepest sternal depression **A ratio of the transverse to AP diameter of the inner chest wall of greater than 3.25 is considered significant for Pectus.** This ratio has been used to select patients for surgical correction



- Lung volumes are usually normal or mildly reduced.
- If a restrictive impairment is present, it is positively associated with the degree of sternal depression and is more severe if there is associated scoliosis.
- The RV may be normal or slightly .  
FEV1 /FVC ratio is typically within normal limits  
FRC is usually within the normal range
- Although deformed, the mobility of the rib cage is not impaired during quiet breathing lung compliance in Pectus excavatum is within normal limits

### **Exercise Capacity**

- Cardiopulmonary exercise testing is often normal . Maximal work rate, oxygen consumption, and heart rate, as well as the oxygen pulse are similar to controls.
- In patients with more severe deformities may have a mild reduction in the maximal work rate or a decrease in the oxygen consumption for a given work.

- This has been attributed to decreased venous return to the heart.
- The depressed sternum may compress the right atrium, reduce cardiac output, and limit exercise.
- In support of this assertion, cardiac anomalies, such as compression of the right ventricle, narrowing of the RVOT and sacculations of the right ventricular wall, have been observed using two-dimensional cross-sectional echocardiography
- However the association with right ventricular dysfunction is uncertain at best.

### Surgical repair of Pectus.

Candidates for surgical repair are patients with a transverse-to-AP diameter ratio of greater than 3.25 as determined by CT of the chest.

The surgical approach to Pectus excavatum has evolved over time

- **The Ravitch repair** included resection of costal cartilage and sternal osteotomy with or without fixation of the sternum with external or internal supports.
  - invasive nature
  - sternal necrosis,
  - infection and
  - recurrence of the deformity, especially in younger children in whom sternal supports are not used.
- Less invasive approaches to Pectus excavatum have been developed. Of these, the **Nuss procedure** is widely used as an alternative
- A customized, curved metal bar under the sternum at the point of its deepest depression through small incisions made on each side of the chest. The bar is removed after 2 to 4 years when permanent chest wall remodeling has occurred
  - minimally invasive approach with
  - immediate cosmetic improvement,
  - shorter operating time,
  - less time to return to regular activities, and
  - excellent long-term results

- Problems include
  - bar displacement or rotation requiring reoperation,
  - pneumothorax,
  - Pericarditis, and
  - Infection
- Surgical repair provides cosmetic benefits and may positively affect the psychosocial well-being of the patient
- The pathophysiologic benefits of the repair remain controversial.
- Exercise capacity and dyspnea often improve, but pulmonary function typically decreases.
- Most studies on pulmonary function in the invasive Ravitch operation.
- Has been attributed to abnormal chest wall mechanics as a result of articular changes in the sternal and parasternal areas.
- It is hypothesized that the less invasive Nuss procedure might have fewer adverse effects on postoperative pulmonary function

## 4.Flail chest

- Flail chest denotes a condition in which fractures of ribs produce a segment of the rib cage that is disconnected from the rest of the chest wall and deforms markedly with breathing.
- Usually, double fractures of three or more contiguous ribs or combined sternal and rib fractures are needed to create the flail segment.
- The detached part of the rib segment is pulled inward with inspiration and bulges outward with expiration compromising ventilation
- Estimated to occur in up to 25% of adults but in only 1% of children following blunt chest trauma.
- This lower incidence in children may be related to their greater chest wall compliance

## Etiology of Flail Chest:

- Trauma:
  - Automobile accidents, falls
  - After cardiopulmonary resuscitation
- Pathologic fractures
  - multiple myeloma,
  - other rib metastases
- Sternectomy and simple rib fractures
- Corrective rib resection
- Congenital

- Flail chest is a clinical diagnosis made by observing the paradoxical motion of the chest wall in the spontaneously breathing patient with blunt trauma and radiographic documentation of rib or sternal fractures.
- Chest radiographs underestimate the presence and extent of rib fractures
  - hemothorax and
  - pulmonary contusion
  - If the patient is intubated and ventilated with positive-pressure ventilation.
- Special rib views or oblique films can diagnose many rib fractures missed by plain chest radiographs.
- Nuclear medicine scans are the most accurate in detecting rib.
- A three-dimensional reconstruction of a thoracic helical-CT scan done to evaluate the extent of thoracic injuries, can also identify rib fractures.

- TLC, VC, and FRC can be reduced to 50% of predicted
- The initial reductions attributed to disordered chest wall motion. VC either returns to its baseline value within 6 months or remains mildly reduced.
- In contrast, patients with pulmonary contusion complicating flail chest may have persistent reductions in FRC for up to 4 years due to fibrous changes in the contused area.

## **Respiratory Mechanics**

- The multiple rib fractures uncouple the flail segment from the remainder of the chest wall.
- During inspiration, the sub-atmospheric pleural pressure is unopposed, and the uncoupled segment of chest wall moves paradoxically inward during inspiration. During expiration, pleural pressure becomes less negative, and the flail segment can be seen moving paradoxically outward.
- A reduction in lung compliance due to pulmonary contusion or microatelectasis or an increase in airway resistance further lowers pleural pressure during inspiration and worsens the flail.



- On the basis of rib fractures and anatomic location of flail segment, three types of flail chest have been described:
  - lateral,
  - anterior, and
  - posterior.

Lateral flail chest is the most common;

The flail segment is located posteriorly and laterally.

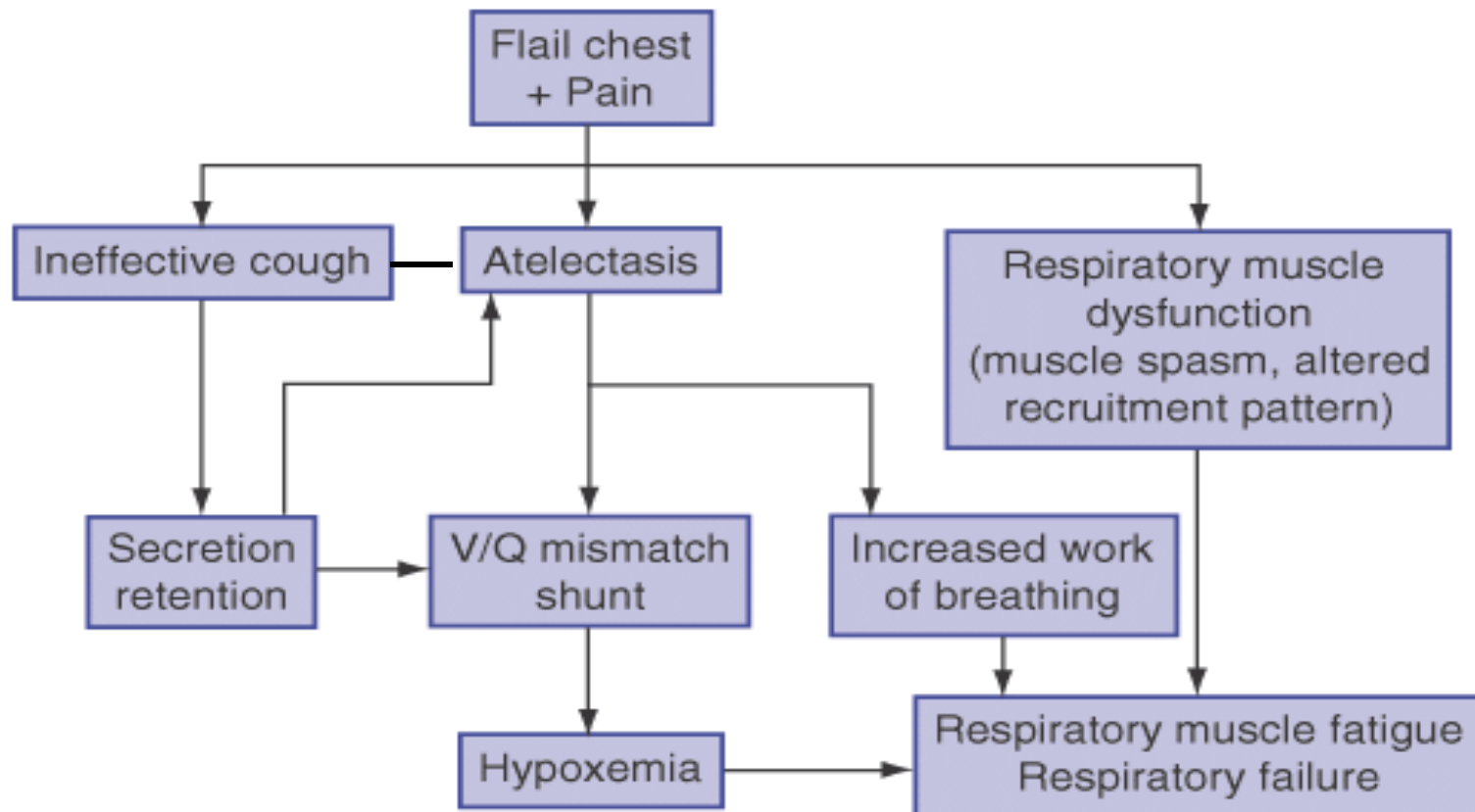
Anterior flail chest occurs when there is separation of the sternum from the ribs.

Posterior flail chest is associated with less severe clinical derangement because of splinting provided by the back muscles.

However, this anatomic classification is merely descriptive and does not provide any information about the actual degree of chest wall distortion in flail chest.

- Chest wall distortion is better characterized by recording the respiratory changes in the dimensions of not only the rib cage but also the abdomen
- Paradoxical motion may occur between the rib cage and abdomen or within the rib cage itself.
- Abnormal patterns seen include
  - (1) inward displacement of the transverse rib cage as the lower anterior rib cage and abdomen are expanding,
  - (2) inward displacement of the anterior lower rib cage and abdomen as the transverse rib cage is expanding, and
  - (3) inward displacement of the lower rib cage as the transverse rib cage and abdomen are expanding.
- A given location of flail segment not necessarily associated with a specific pattern of distortion
- These different flail patterns may therefore reflect not only differences in the location of the rib cage fractures but also differences in respiratory muscle recruitment patterns.

# Pathophysiology of flail chest



- Initially, respiratory failure was attributed to *pendelluft*
- Clinical observations and recent experimental data suggest that the pathogenesis is complex
- Flail chest and the pain that invariably accompanies chest trauma lead to
  - cough impairment,
  - regional atelectasis,
  - rib cage muscle spasm, and
  - specific changes in the pattern of muscle activation and recruitment.
  -
- Paradoxical motion of the flail segment may also increase the degree of muscle shortening during inspiration adding to energetic cost.
- Inspiratory muscles become inefficient when operating over shorter lengths the paradoxical motion of the flail segment may result in diaphragm inefficiency

- Pulmonary contusion or pleural restriction add to the work of breathing by increasing the negative pressure needed to inhale.
- The added work of breathing, along with respiratory muscle inefficiency increases the oxygen cost of breathing and predisposes these muscles to fatigue
- Atelectasis and lung contusion in conjunction with respiratory muscle fatigue result in
  - hypoxemia,
  - hypercapnia, and
  - ultimately respiratory failure.
- Hypoxemia due to either process would contribute to respiratory muscle dysfunction by reducing energy supplies to the muscles.
- The presence of multiple vicious cycles ensures perpetuation of the process.

## Implications of Flail Chest:

- Traumatic flail chest is a marker of severe chest injury
- An independent risk factor for significant respiratory complications, respiratory failure, and death
- Flail chest promotes atelectasis. Atelectasis, via its effect on compliance, increases the severity of flail chest and lead to worsening of respiratory failure.
- Patients with isolated flail chest developed respiratory failure and required mechanical ventilation almost twice as often (57%) as did patients with isolated pulmonary contusion (31%).
- The mortality rate from chest wall trauma alone ranges from 7% to 14%.
- In a series of 1026 multiple-trauma patients, the mortality rate for those with flail chest was 68%, compared with a 27% mortality rate for the entire group attributable to other associated injuries.
- A significant correlate of survival in patients with traumatic flail chest is the presence or absence of head injury

- The lung may also be injured in patients who sustain chest wall trauma.
- Pulmonary contusion, hemothorax, and pneumothorax, can occur in up to 60% of patients with flail chest
- Patients with multiple trauma and lung contusion had a mortality rate (56%) almost as high as those with flail chest (67%).
- Pulmonary contusion is best detected by chest CT scans. If the contusion is seen on the chest radiograph, however, it may be more clinically significant.
- If the patient survives the initial injury, symptoms of chest tightness, chest pain, or dyspnea on exertion is seen in half of patients. Operative stabilization of the chronic flail chest can correct the defect and dramatically relieve the symptoms.
- Because of an incompetent chest wall in patients with flail chest, external chest compressions during cardiopulmonary resuscitation are ineffective. In such cases, internal cardiac massage may be recommended

## TREATMENT-General Aspects

- The primary goal of treatment is to restore the anatomic and functional integrity of the chest wall by providing stability of the flail segment.
- This can be accomplished with conservative management or with surgical intervention.
- Pain should be adequately controlled in all patients with flail chest.
- Chest wall pain may cause patients to adopt a rapid, shallow breathing pattern, worsening atelectasis and lead to arteriovenous shunting and hypoxemia.
- Analgesia
  - reduces splinting,
  - improves tidal volume, and
  - facilitates cough.
- Pain control can be accomplished by
  - oral medications,
  - patient-controlled analgesia pumps,
  - intermittent intercostal nerve blocks, or
  - epidural anesthesia.



- Mechanical ventilation has been used in the treatment of flail chest for more than 40 years.
- Such "internal pneumatic stabilization" consisted of tracheostomy with a prolonged 3- to 5-week period of mechanical ventilation.
- Mechanical ventilation failed to improve survival and predisposed patients to several complications such as
  - pneumonia,
  - barotrauma,
  - sepsis, and
  - tracheal stenosis.
- Is no longer recommended as a primary means of stabilizing the chest wall; recommended when
  - there is concomitant central nervous system or intra-abdominal injury,
  - shock, or
  - a need to operate for other injuries.
- The role of noninvasive mechanical ventilation in stabilizing the flail segment has not been fully evaluated in patients with flail chest.

- When mechanical ventilation is needed to treat respiratory failure, the mode of ventilation chosen should minimize the resistive and elastic loads imposed by the ventilator.
- IMV has been frequently used as a mode of ventilating patients with flail chest
- Spontaneous breaths through the added resistance of the IMV circuit may aggravate chest wall distortion.
- low impedance modes such as CPAP are accompanied by less chest wall distortion during inspiration. Reduce the risk for Inspiratory muscle fatigue and may be preferred for weaning.
- Supported by a RCT in which patients treated with mask CPAP alone have fewer complications, fewer hospital and intensive care unit (ICU) days, and less ventilator time than patients with similar degrees of blunt thoracic trauma treated with assist-control ventilation.
- Pressure support ventilation may be an alternate means of reducing the ventilator-imposed loads, but the effects of this mode on chest wall distortion have not been evaluated

## Surgical management:

- **Purpose of surgical stabilization is to achieve a mechanically stable chest**
  - to reduce ventilator time
  - avoid ventilator-associated complications.
- **Initial attempts consisted of applying tape, strapping, and external devices to the chest wall. These attempts met with limited success**
- **By externally stabilizing the chest wall, inward displacement of the ribs during the healing phase can be avoided, and the chest wall contour can be restored to normal. No randomized studies available & effectiveness of this modality not defined**
- **Possible indications include**
  - Ventilator-dependent patients able to protect their upper airways with no need prolonged mechanical ventilation.
  - flail chest patients undergoing thoracotomy for intrathoracic injuries.
  - young patients with severe chest wall deformation and patients with large, unstable segments ("stove-in chest")
  - Patients with borderline pulmonary function may be candidates for external fixation.
- **Chest wall external fixation in such patients**
  - improves respiratory mechanics,
  - reduces the duration of mechanical ventilation,
  - reduces hospital stay
  - decreases the incidence of pulmonary infection and
  - barotrauma

## Obesity & The Lung

- Obesity constitutes a significant health problem worldwide. In the United States, it is estimated that more than half of all adults are overweight or obese. It is an increasing problem in developing countries.
- Its prevalence continues to increase. In the U.S, an increase of more than 25% over the last three decades was seen. The number of individuals with severe obesity has tripled
- It is a leading cause of preventable death in the United States, second only to cigarette smoking
- Morbidity and mortality associated with obesity increase with increasing BMI.
- Intra-abdominal fat distribution is also an independent risk for cardiovascular disease and mortality. Upper body or central fat distribution has a greater effect on pulmonary function than lower body fat distribution. Furthermore, abdominal fat distribution is more often associated with sleep-disordered breathing
- World Health Organization and the National Institutes of Health have classified obesity as follows: based on the BMI
  - 18.5- 24.9 kg/m<sup>2</sup> normal
  - 25.0 -29.9 kg/m<sup>2</sup> overweight
  - 30.0 -34.9 kg/m<sup>2</sup> Grade 1 obesity.
  - 35.0-39.9 kg/m<sup>2</sup> Grade.2 obesity
  - >40.0 kg/m<sup>2</sup> Grade 3(morbid) obesity

## Body Composition in Obesity

<i>Parameter</i>	<i>Normal</i>	<i>Mild Obesity</i>	<i>Severe Obesity</i>
BMI(kg/m <sup>2</sup> )	23	34	50
Body fat (%)	22	35	45
Body weight (kg)	65	96	132
Fat-free mass (kg)	51	63	72
Body cell mass (kg)	27	32	36
Total body water (L)	38	44	49
REE (watts)	68	86	96

- Obesity can be evaluated in three contexts:
  - simple obesity, in which individuals are eucapnic with no pulmonary compromise;
  - morbid obesity, in which pulmonary complications are present;
  - obesity hypoventilation ( $\pm$ OSA) in which there is morbid obesity with carbon dioxide retention during wakefulness.

### **Pulmonary Function**

The ERV is reduced, on average, to 60% of the predicted due to the effects of abdominal contents on the position of the diaphragm. .

VC and TLC are usually normal in simple obesity but may be reduced by 20% to 30% in morbid obesity

In general, RV is not reduced in simple obesity. Consequently, the RV/TLC ratio may be greater than normal when obesity causes restriction.

The FEV1 /FVC ratio is usually normal

MVV and peak inspiratory flow rate are also normal, provided respiratory muscle strength is normal

- Spirometry in individuals with obesity hypoventilation syndrome (OHS) usually reveals a more severe restrictive pattern with reductions in
  - TLC, VC,
  - MVV, and
  - peak inspiratory flow rate
- This appears to be related to weakness of the respiratory muscles.
- The ERV may be reduced to 35% of predicted and the FVC to 60% of predicted
- The adverse effects of obesity on pulmonary function cannot be entirely explained by the absolute load of adipose tissue on the chest wall because similar degrees of obesity in simple obesity and OHS result in different patterns of lung volume changes .
- The single-breath diffusion capacity is usually normal or increased in simple obesity and slightly reduced in OHS

- **OHS>Obesity generally lowers compliance of the lung, chest wall, and total respiratory system.**
  - due to weight pressing on the thorax and abdomen thereby imposing an elastic load.
  - threshold-type load on the chest wall in which pleural pressure must be lowered to a sufficient degree before inspiratory flow can begin.
- Specific airway conductance may be reduced to 50% to 70% of normal.
- **FEV1 /FVC ratio is normal**
  - increased airway resistance in obesity appears to lie in lung tissue and small airways
  - Impediment to breathing in obesity results from a two- to threefold increase in intra-abdominal pressure
- **The transition from upright to the supine position reduces lung volume due to increased intra-abdominal pressure & promotes expiratory flow limitation during tidal breathing**
- **The work of breathing is 60% higher in simple obesity and may be as much as 250% higher in OHS with an Increase in the energy cost of breathing; the oxygen cost of breathing at rest**
- **This places obese patients at risk for respiratory failure during conditions characterized by increased ventilatory demands such as an intercurrent illness.**



# Gas Exchange

- Hypoxemia may be mild or absent in simple obesity. The arterial PO<sub>2</sub> is even lower in OHS.
  - Ventilation-perfusion mismatch results from airway closure
  - in OHS venous admixture and
  - hypoventilation may also contribute.

## Control of Breathing

In obese eucapnic individuals, the resting ventilation and the respiratory drive are either normal or increased when compared with those of non obese subjects as are the ventilatory responses to hypoxia and hypercapnia

In patients with OHS, the ventilatory drive is inappropriately low.

The ventilatory response to hypoxia is also decreased, frequently to a greater extent than the hypercapnic response.

The etiology of the attenuated ventilatory drive in patients with OHS is not fully known

## NIV in obesity

- CPAP for patients with OSA.
- BiPaP for
  - Obese patients who experience nocturnal or nocturnal and daytime hypercapnia.
  - OSA patients not tolerating high CPAP pressures required
  - OSA patients having obstruction despite high CPAP pressures.
- BiPaP has been used successfully to treat atelectasis after gastroplasty
- Positive-pressure ventilation
  - improves sleep quality, *(CHEST 2001; 119:1102–1107)*
  - corrects hypercapnia, and
  - improves daytime oxygenation