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Foreword

Greetings from the Andhra Hospitals!

For the past two decades, we are delivering excellence in terms of quality health care in the field of Paediatrics including Paediatric and Neonatal intensive care, Paediatric cardiac intensive care and other allied Paediatric sub-specialities.We take pride in introducing ECMO in pediatric specialty for the first time in the state of Andhra Pradesh. In this context, we convey our heartfelt thanks to you for your continued support and encouragement which played a pivotal role in our success.

As a token of our commitment to contribute to continuous medical education, we are introducing this monthly E-journal to showcase important clinical guidelines, recent advances in paediatric sub-specialties, interesting case reports, image quiz, OSCE scenarios etc, gathered from our patient database.

We hope this endeavor would prove to be useful to practicing paediatricians, intensivists, neonatologists and post-graduate students.

Please send your valuable feedback and suggestions to ***maramkp@gmail.com***.

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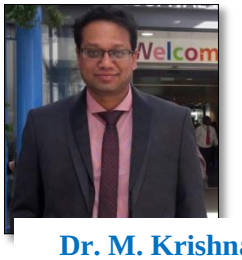

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Index

ARTERIAL BLOOD GAS ANALYSIS

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Introduction

The most effective method for assessing the patient's gas exchange is by measuring the values of oxygen (O_2), carbon dioxide (CO_2) and pH of the arterial blood (Hazinski, M 1999). An ABG analysis evaluates how effectively the lungs are delivering oxygen to the blood and how efficiently they are eliminating carbon dioxide from it. The test also indicates how well the lungs and kidneys are interacting to maintain normal blood pH (acid-base balance). Blood gas studies are usually done to assess respiratory disease and other conditions that may affect the lungs, and to manage patients receiving oxygen therapy. In addition, the acid-base component of the test provides information on renal function.

Defining terms

pH: A reflection of the blood hydrogen ion (H+) concentration. Normal pH =7.4 (Range 7.35 – 7.45). pH is calculated on a logarithmic scale.

PaO_2: partial pressure of oxygen dissolved in the blood

$PaCO_2$: partial pressure of dissolved CO_2 in the blood

Bicarbonate (HCO_3^-): Bicarbonate is regulated by the lungs (through CO_2 removal) and by the kidneys (through H^+ ion and HCO_3^- excretions or reabsorbtion). Normal HCO_3^- is 22 – 28 mEq/ml

Base deficit: a calculated number (by the gas machine) that represents the theoretical deficit (of a base) or excess (of an acid) present in the sample. A base deficit indicates metabolic acidosis. (< -2)

Base excess: A positive value represents the theoretical excess of a base and this indicates metabolic alkalosis. (> +2).Normal base deficit/excess = -2 to +2

Acidosis: Too much acid in the blood – pH < 7.35

Alkalosis: Too much alkali (or base) in the blood – pH > 7.45

Hypoxaemia: Low level of oxygen in the blood, generally considered to be PaO_2 < 75 mmHg when the patient is breathing room air (21% oxygen) at sea level.

Hypercarbia/hypercapnia: High level of CO_2 in the blood with a $PaCO_2$ > 45 mmHg

The Method.

The blood gas results should be considered in parts. The result below shows what each part represents.

RESULT	PART	REPRESENTS
pH	Acid-base	pH scale – acidity to alkalinity
$PaCO_2$	Respiratory indicator	Potential cause of the pH problem.
HCO_3^- std	Metabolic indicator	Potential cause of the pH problem.

The pH result presents us with a potential acid-base problem. The $PaCO_2$ and the HCO_3^- present us with possible causes for the problem. The first thing to do is to identify the problem and then determine what has caused it.

Step 1 : Identify the source.

The first thing to do is to identify whether the blood sample has been obtained from a capillary, a venous (central line) or an arterial source. It will affect the interpretation and applicability of the blood gas results.

1. Arterial samples provide the most accurate results.
2. Significant changes occur to the oxygen_content of the blood (PaO_2) as it passes from the artery through the capillary bed.
    - A wide range of oxygen values is possible when the source comes from a capillary stab.

- The degree of squeezing that went into providing sufficient capillary blood for the sample also affects the oxygen result.
- Venous blood is usually about 70% saturated and has a PaO_2 of about 40 mmHg but the value does not correlate well with arterial oxygen content

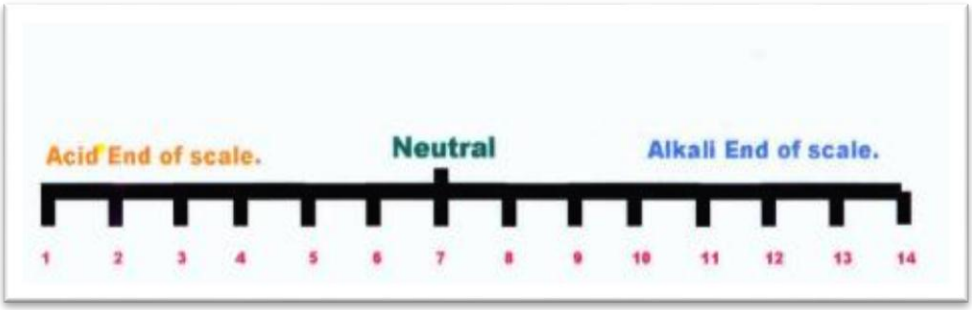
3. **The pH of venous or capillary blood is usually about 0.033 lower than in arterial blood.** It has slightly greater acidity. This difference is not big enough to make a difference to decisions on patient care.
4. **The carbon dioxide level ($PaCO_2$) of capillary or venous blood is usually about 4.5 mmHg higher than in arterial blood**. This difference is big enough to make a difference to blood gas interpretation. The effect of this difference on the $PaCO_2$ interpretation will be discussed in the relevant section.

Step 2: Identify the pH part of the gas

Normal limits	
pH	**7.35 – 7.45**

The above values apply to arterial, capillary and central line samples.

- pH is measured on a scale from 0 – 14. Low values represent the most acid and high values representing the most alkali. pH 7 is neutral : this is the pH of sterile water. A diagram of a pH scale is printed below. You can see the acid and the alkali ends of the scale marked.The pH scale represents the hydrogen ion concentration as a negative logarithm. As the hydrogen ion concentration increases, the pH number falls. A change in pH from 7.4 to 7.35 represents a much larger increase in the hydrogen ion concentration than the change from 7.45 to 7.35.



- The normal pH of blood occupies a very small range on a scale from one to 14.
    - Adult stomach acid has a pH of 1 – 4.
    - Normal urine has a pH ranging from 5.5 – 7 depending on the type of diet that is eaten.
    - Bile has a pH of 7.8 – 8.8.
- The above diagram shows the part of the pH scale between 6.0 and 8.0. This is the range into which most of the blood gas pH values seen in PICU will fall. The body can cope for some time with a blood pH which falls outside the reference range of 7.35 – 7.45. This scale shows the sort of pH which the body can tolerate. If the blood pH is deranged to the extent that it falls outside this range, then the patient is either already dead or likely to die imminently.
- How severe is the pH problem?
- It is vital that the blood pH is maintained within this narrow range. A pH of 7.2 or as high as 7.6 is tolerable but still needs correction in most cases. (See permissive hypercapnea on the $PaCO_2$ page for the exception to this rule). Cell destruction commences outside these ranges and patients will die if their pH falls and remains below this level. Once the severity of the pH problem is established, two categories of severity have been suggested. The first is 'severe' and the second is 'life-threatening'.

When assessing the pH, three questions need to be asked.

1. Is the pH within normal limits?
2. If the pH is not within normal limits, is it showing an acidosis or an alkalosis?

3. If there is an alkalosis or an acidosis, how severe is the problem?

If the pH is not within normal limits, is it showing an acidosis or an alkalosis?

The term acidotic is used for any blood pH less than 7.35

- The term alkalotic is used for any pH value greater than 7.45.

You should now be able to identify whether the pH is within normal limits and whether any pH problem is 'acidosis' or 'alkalosis'. If you don't feel able to do this, read back over the previous section.

1. "Severe" is used for
    - any acidosis, where the pH is less than 7.2 and greater than 7.0
    - any alkalosis where the pH is greater than 7.6 and less than 7.8.
2. "Life-threatening"is used for
    - any acidosis, where the pH is less than 7.0
    - any alkalosis where the pH is greater than 7.8.

If the pH result shows a "severe" problem, action needs to be taken promptly. If the pH result shows a "life-threatening" problem, action needs to be taken urgently to try to prevent an arrest.

The rest of the gas result will show the cause of the acidosis or alkalosis. Once the cause has been identified, appropriate corrective action can be taken below 7.0 or above 7.8.

Step 3 : Assess the respiratory part of the gas : the $PaCO_2$

From this point on, it will probably be easier if a practical example is used.

Look at the gas result below and decide what note you would make to yourself about the **pH**. Look at each value and tick whether the result is normal or abnormal.

	Gas Result	Normal	Abnormal
pH	7.29		
$PaCO_2$	52 mmHg		
HCO_3^- std	25		

The respiratory part of the blood gas comprises two results:

- The carbon dioxide level,
  - $PaCO_2$ (partial pressure of carbon dioxide)
- The oxygen level,
  - PaO_2 (partial pressure of oxygen).

There are two questions that need to be asked about the carbon dioxide level for blood gas interpretation.

1. Is the carbon dioxide level ($PaCO_2$) within normal limits?
2. Would this result cause acidosis or alkalosis?

CO_2 reacts with water in the body to form carbonic acid, which then breaks down to form bicarbonate and hydrogen ions. The higher the level of CO_2, the more acidic the patient will become

$H_2O + CO_2 \rightarrow\leftarrow H_2CO_3$ (carbonic acid) $\rightarrow\leftarrow HCO_3^- + H^+$

Is the carbon dioxide level ($PaCO_2$) within normal limits?

Normal arterial limits	**Normal capillary/venous limits**
35 – 45 mmHg	**40 – 50 mmHg**

If the sample is from a central line (venous) or capillary source, the pCO_2 is normally 4.5mmHg higher than the arterial sample

Would the $PaCo_2$ result cause acidosis or alkalosis?

$PaCO_2$	**pH**	**Result**
High	Acidosis.	Respiratory acidosis.
Low	Alkalosis.	Respiratory alkalosis.

A high carbon dioxide level means greater than average concentrations of carbonic acid in the blood.

- If the pH of the blood is acidotic and the carbon dioxide level is high, the carbonic acid would be the cause of the problem. This would be a 'respiratory acidosis'.
- Hypo-ventilation problems are the cause of 'respiratory' acidosis. The patient's ventilator settings will need adjusting. If the patient is not ventilated, he/she may need intubation and ventilation.
- Respiratory acidosis is the most common problem seen on general PICU.

A low carbon dioxide level indicates less than the usual amount of carbonic acid circulating.

- If the pH of the blood is alkalotic and the carbon dioxide is low, insufficient carbon dioxide would be the cause. This would be a 'respiratory alkalosis'.
- Hyper-ventilation is the cause of 'respiratory' alkalosis. The patient's ventilator settings will need adjusting. Hyperventilation is rare in non-intubated patients, but can occur as a response to stress and anxiety.

$PaCO_2$	pH	Result
High	Acidosis.	Respiratory acidosis.
Low	Alkalosis.	Respiratory alkalosis.

If the $PaCO_2$ is normal, then we need to look for some other cause of the deranged pH.

On the practice example, tick whether **the $PaCO_2$** level is normal or abnormal

	Gas Result.	Normal	Abnormal
pH	7.29		
$PaCO_2$	52 mmHg:		
HCO_3^- std	25		

A cause for the acidosis in this example has now been found. Even if the $PaCO_2$ value has not caused the pH problem, the identification of a deranged PaCO2 made by the physician will be helpful when more complex situations need to be interpreted (Mixed causes and compensations.)

Step 4 : Assessing the metabolic part of the gas : the Standard Bicarbonate.

The standard bicarbonate (HCO_3^- std) and the base excess represent the 'metabolic' part of the gas result.

Metabolism involves:

- Anabolism - the building up of substances necessary for life.
- Catabolism - the breaking down of substances necessary for life.

As a result of metabolic processes, a number of acids are produced. These are normally excreted safely without causing anything but temporary problems of pH adjustment. In disease processes, the acids may not be excreted, or may be produced in excess, so that an elevated level of these metabolic acids persists in the bloodstream.

Consider the following two disease processes and their effect on blood acidity:

1. **In hypoxic states**, the metabolism of glucose to energy, carbon dioxide and water cannot be fully completed. Instead of carbon dioxide being formed, lactic acid is produced. If the hypoxia is not resolved, the level of lactic acid builds up and lowers the pH.

Increasing lactate levels (as noted on a blood gas) can indicate poor systemic perfusion or hypoxia, e.g. in sepsis, burns, severe hypoxia, post-op cardiac patients, hypovolaemia

2. In health, insulin allows glucose to enter the cell. **In diabetic keto-acidosis,** glucose cannot enter the cell to provide fuel for energy production, because of a shortage of insulin. The cells are devoid of glucose and send out chemical signals that more glucose is needed. The body begins to break down fats to provide more glucose for the cells. A vicious cycle of increasing blood glucose and increasing fat breakdown ensues. Ketones are a product of fat breakdown and cause the keto-acidosis.

These are just two of the possible metabolic acid culprits for lowering the pH. Measuring all the possible acid causes individually would be a lengthy process. Instead, the blood gas machine measures the substance that the body uses to 'mop up' hydrogen ions: Bicarbonate (HCO_3^-).

Bicarbonate is an important buffer which mops up carbonic acid from respiratory acidosis (i.e. it attempts to neutralise the acid). There are two bicarbonate results recorded on the blood gas – standard and actual. Standard bicarbonate means the bicarbonate level when it is measured under standard conditions, so only the effect

of metabolic acids will be detected. Therefore, this is the one we record and interpret. In the blood gas machine, standard conditions for this measurement are:

PaCO2 40 mmHg Temp $37°C$

The above conditions mimic normal healthy respiratory status, with a pCO_2 of 40 mmHg and normal patient body temperature.

Let's consider a new blood gas. Please tick what is normal and abnormal:

	Gas Result	Normal	Abnormal
pH	7.29		
$PaCO_2$	44 mmHg		
HCO_3^- std	18 mmol/L		

When considering the HCO_3^- std, two questions need to be asked.

1. ***Is the standard bicarbonate level (HCO_3^- std) within normal limits?***
2. ***Would this result cause acidosis or alkalosis?***

Is the standard bicarbonate level (HCO_3^- std) within normal limits?

Normal limits.	
Standard Bicarbonate - HCO_3^- std	22-26 mmol/L

Would this result cause acidosis or alkalosis…….?

- If there is an excess of bicarbonate, there will not be much acid. It would cause an alkalosis: metabolic alkalosis.

- If there is a deficiency of bicarbonate, there is more acid than the bicarbonate can deal with. It would cause an acidosis: metabolic acidosis.

HCO_3^- std	pH	Result
Low	Acidosis	Metabolic acidosis.
High	Alkalosis	Metabolic alkalosis.

The most likely cause of metabolic alkalosis is chronic lung disease (CLD) or over-administration of sodium bicarbonate or another buffer solution. It may also be seen in some cases of poisoning.

The following table shows all the blood gas interpretations which you have learnt to make so far.

Acid Base	PH	$PaCO_2$	Bicarb
Acute Respiratory Acidosis (uncompensated)	< 7.35 ↓	↑ >45	Normal
Respiratory Acidosis (Partial compensation)	< 7.35 ↓	↑ >45	↑ >26
Chronic Respiratory Acidosis (compensated)	Normal	↑	↑
Acute Respiratory Alkalosis (uncompensated)	↑	↓	Normal
Respiratory Alkalosis (Partial compensation)	↑	↓	↓
Chronic Respiratory Alkalosis (compensated)	Normal	↓	↓

Acid Base	PH	$PaCO_2$	Bicarb
Acute Metabolic Acidosis (uncompensated)	< 7.35 ↓	Normal	↓ <22
Metabolic Acidosis (Partial compensation)	< 7.35 ↓	↓ <35	↓ <22
Chronic Metabolic Acidosis (compensated)	Normal	↓ <35	↓ <22
Acute Metabolic Alkalosis (uncompensated)	↑	Normal	↑ >26
Metabolic Alkalosis (Partial compensation)	↑	↑ >45	↑ >26
Chronic Metabolic Alkalosis (compensated)	Normal	↑ >45	↑ >26

Dealing with pH problems involves trying to correct the factor that is causing the pH problem and maintain life, while the systems of the body recover from disease.

Methods for doing this are:

Problem	Causes of problem	Methods of dealing with the problem
Respiratory acidosis.	• Lung disease. • Respiratory depression - due to drugs or head	• Intubate and ventilate - if not intubated. • Increase minute volume by
Respiratory	• Over-ventilation on the	Reduce the rate and/or tidal volume.

Metabolic acidosis.	• Increased lactic acid due to poor oxygenation. • Diabetic acidosis. • Renal failure.	Treat the cause firstly then give buffer: 1. Sodium bicarbonate. 2. H.A.S. 4.5% or other blood products as needed The
Metabolic alkalosis.	• Over administration of sodium bicarbonate. • Poisoning or overdose	Do not give bicarbonate without knowing the HCO_3^- std value.

Step 5 : The Base Excess or Base Deficit.

Normal limits.	
Base excess	-2 to +2 mmol/L

It expresses the difference between the **patient's** standard bicarbonate level and the **normal** standard bicarbonate (24 mmol/L).

- If the HCO_3^- std is 30 mmol/L, that is 6 mmol/L more than 24 mmol/L. The patient has a base excess of +6 mmol/L.
- If the HCO_3^- std is 20 mmol/L, that is 4 mmol/L less than 24 mmol/L. The patient has a base excess of -4 mmol/L or a base deficit of 4 mmol/L.

A base excess between……

- -2 mmol/L (representing a HCO_3^- std of 22 mmol/L) and
- +2 mmol/L (representing a HCO_3^- std of 26 mmol/L)

…….is considered to be within the normal range.

Tick the appropriate boxes and calculate the **Base Excess** for the following example:

	Gas Result	Normal	Abnormal
pH	7.29		
$PaCO_2$	52 mmHg		
HCO_3^- std	27 mmol/L		
Base excess			

Step 6 : Check for compensation.

What is compensation?

In the earlier section on pH, categories of pH result were established. The categories were 'life-threatening' or 'severe'. In the same way that nurses and medical staff would wish to correct these problems promptly, so also does the body. It tries to establish a normal pH again. This is **compensation.** (N.B. before compensation mechanisms take place gases are considered uncompensated)

There are four ways the body can do this:

1. If the problem is acidosis, it can:
    - get rid of more carbon dioxide (take deeper breaths or breathe faster)
    - retain/produce more bicarbonate to neutralise the acid (via the renal system)
2. If the problem is alkalosis, the body can:
    - Retain more carbon dioxide (take shallower breaths or breathe out less often).
    - Lose more bicarbonate (via the renal system).
    - (Remember that alkalosis is a much less common problem than acidosis, so you will not see many alkalotic problems on the ICU.)

Obviously if one of the respiratory or renal systems is not functioning properly because of illness or injury, the body will be more dependent on the other method for excreting waste acids.

There are two types of compensation.

1. Partially compensated, where the pH, pCO_2 and HCO_3^- are all abnormal but either the $PaCO_2$ or Bicarbonate are trying to compensate (normalise) the pH
2. Compensated – where the pH is within normal limits, but the pCO_2 and HCO_3^- std are abnormal

	Gas Result	**Notes.**
pH	7.29	*Acidotic*
$PaCO_2$	52 mmHg	*High $PaCO_2$. Would cause acidosis*
HCO_3^- std	27 mmol/L	*High HCO_3^- std. Would cause alkalosis*
Base excess	+3 mmol/L	

Consider the example gas again.

1. A respiratory acidosis can be identified.
2. There are, however, three abnormal results and the $PaCO_2$ and HCO_3^- std would cause different problems.
    - this is a 'partially compensated respiratory acidosis'.

As in the previous example, what would you write for the gas below?

	Gas Result	**Notes**
pH	7.36	
$PaCO_2$	62 mmHg	

HCO_3^- std	28 mmol/L	
Base excess	+4 mmol/L	

This gas shows compensated respiratory acidosis

The pH is normal, so on PICU we would refer to the blood gas as normal. However, in compensated problems, there has been an acidosis or alkalosis to which the body has reacted to achieve a pH of 7.35 – 7.45.

How can you tell which problem came first?

- Did the acidosis come first and then the body compensated to bring the pH back to normal?
- Did the alkalosis come first and then the body compensated to bring the pH back to normal.

The key to working out which problem came first, is to re-examine the pH value. The body never overcompensates.

- If a fully compensated 'acidosis' exists, then the pH will be normal but less than 7.4.
- If a fully compensated 'alkalosis' exists then the pH will be normal but greater than 7.4.

	Gas Result.	**Notes.**
pH	7.36	*normal*
$PaCO_2$	62 mmHg	*High pCO_2. Would cause acidosis.*
HCO_3^- std	28 mmol/L	*High HCO_3^- std. Would cause alkalosis.*

Base excess	+4 mmol/L	

Look at this blood gas which you have just worked on again and follow through the reasoning.

- The pH is normal.
- The $PaCO_2$ would cause acidosis.
- The HCO_3^- std would cause alkalosis.
- The acidosis came first because the pH is less than 7.4.
- This is a compensated respiratory acidosis.

Step 7 : Assessing the oxygen level : PaO_2

Prompt delivery of high concentration oxygen is vital in emergencies.

- If there is not enough oxygen for metabolic processes to completely convert glucose to carbon dioxide, water and energy, lactic acid builds up. This could lower the pH to dangerous levels.
- The brain also needs a sufficient oxygen supply to continue the process of maintaining all body functions. Deprived of oxygen for more than three minutes under normal temperature conditions, severe cell damage will occur within the brain.

However, oxygen is toxic in overdose.

- High PaO_2 causes retinopathy of prematurity *in preterm babies*.
- Exposure to high levels of oxygen (>60%) for periods even over just 24 hours will damage the lungs and cause nitrogen washout.
- High PaO_2 can cause tissue damage and may also cause cancers due to free radical formation.

When considering the PaO_2, there are number of questions one should ask:

1. Is the oxygen within normal limits?

2. Is the patient's haemoglobin level satisfactory?
3. What age category does the patient fall into ;
    - Neonate or preterm infant.
    - Older child.
4. Would the source of the sample make this PaO_2 unreliable?
5. Does the patient have a duct-dependant heart lesion or a right-to-left shunt? – These children will have expected lower PaO_2 levels and clinicians tend to go by the oxygen saturations.
6. Does the patient have a cardiac lesion producing unbalanced pulmonary or systemic blood flow (pre-operative HLHS, Truncus Arteriosus, etc) where oxygen administration (by lowering the pulmonary vascular resistance) may be dangerous?

Normal arterial PaO_2 limits	
Patient age	
Older child or adult	75 – 100 mmHg
Neonate or preterm	60 – 80 mmHg

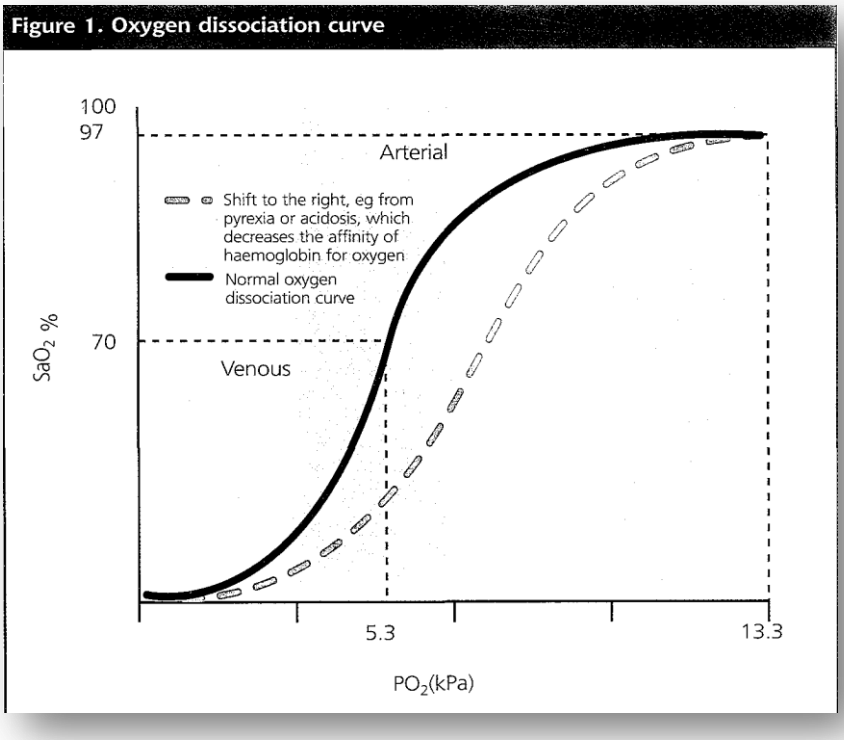
- If the PaO_2 exceeds the recommended amount, weaning of the inspired oxygen can be undertaken.
- If the PaO_2 is low, then the inspired oxygen level should be increased.

However, low PaO_2 coupled with high saturations can be deceptive in patients who are anaemic.
Oxygen is carried round the body attached to the haemoglobin on the red blood cell. If the patient has fewer red blood cells than necessary, the red blood cells can all be fully saturated (carrying their maximum load of oxygen, i.e. 100% saturations) but the overall oxygen content of the blood, the PaO_2, would be low. It is advisable to check that the patient's haemoglobin level is adequate, in cases where a PaO_2 value is low, but the saturations are high. These patients need a blood transfusion.

Why is a lower level acceptable for neonates?

Fetal blood has an even higher affinity for oxygen than the adult type, so it greedily grabs it up from the maternal blood by diffusion at the placenta. The blood then travels along the umbilical cord. Inside the foetus some gets side-tracked through the liver. The liver is a massive chemical factory for the human body and much of the oxygen is used here to supply energy for liver metabolism. Then blood which has not passed through the liver is mixed up with the venous blood returning from the foetal circulation. The PaO_2 level as the blood passes out into the foetal arterial system through the ductus arteriosus or the foramen ovale (by-passing the inactive lungs) is about 30mmHg, yet it keeps the foetal body going! Small babies are probably better at coping with lower oxygen levels than children and adults, because they have been doing it all their intrauterine life. However no-one would recommend letting levels get that low outside the womb.

Oxygen



- Blood oxygen levels fall in the capillary bed as the oxygen passes from the haemoglobin to the tissues. The PaO_2 value can vary considerably. Once the

desaturated blood has entered the vein, it usually has a value of around 40 mmHg.

- Capillary sample values are also subject to error because the tissues are often squeezed in order to obtain a sample.

There is not a reliable correlation between arterial PaO_2 and the PaO_2 from other sources. Physician must rely on saturation monitoring to govern their management of oxygen therapy in such cases. Obviously if the PaO_2 of a capillary or central line sample is very high, it will indicate that the patient is receiving too much oxygen.

If the patient has a **duct-dependent** heart lesion, it is inadvisable to increase their inspired oxygen level without medical instruction. High oxygen levels promote closure of the duct and prevent systemic oxygenation altogether.

Remember that if you have a child with a right to left shunt, their PaO_2 will be low because they have mixing of venous and arterial blood, not because of lung disease. Advice from the cardiology team on a recommended arterial PaO_2 is required.

Finally it must be added that sometimes you just have to be happy with what you can achieve regarding the PaO_2! There is not much one can do to correct the PaO_2 in children who are very sick and receiving greater than 90% inspired oxygen already. (High Frequency oscillation ventilation (HFOV) may be considered to clear carbon dioxide in these patients and Extra-Corporeal Membrane Oxygenation (ECMO) may be considered to improve oxygenation.) The priority in such cases is to try to keep the patient alive by appropriate pH and $PaCO_2$ management until their lungs and their oxygenation improve.

Beware of ventilating any patient on 100% oxygen for prolonged periods. Some nitrogen is vital in the mixture to help keep the alveoli open. If maximum dose oxygen (100%) is required for prolonged periods, then 95% inspired oxygen should be used. The patient will need to be reviewed by the medical staff and an acceptable saturation level discussed.

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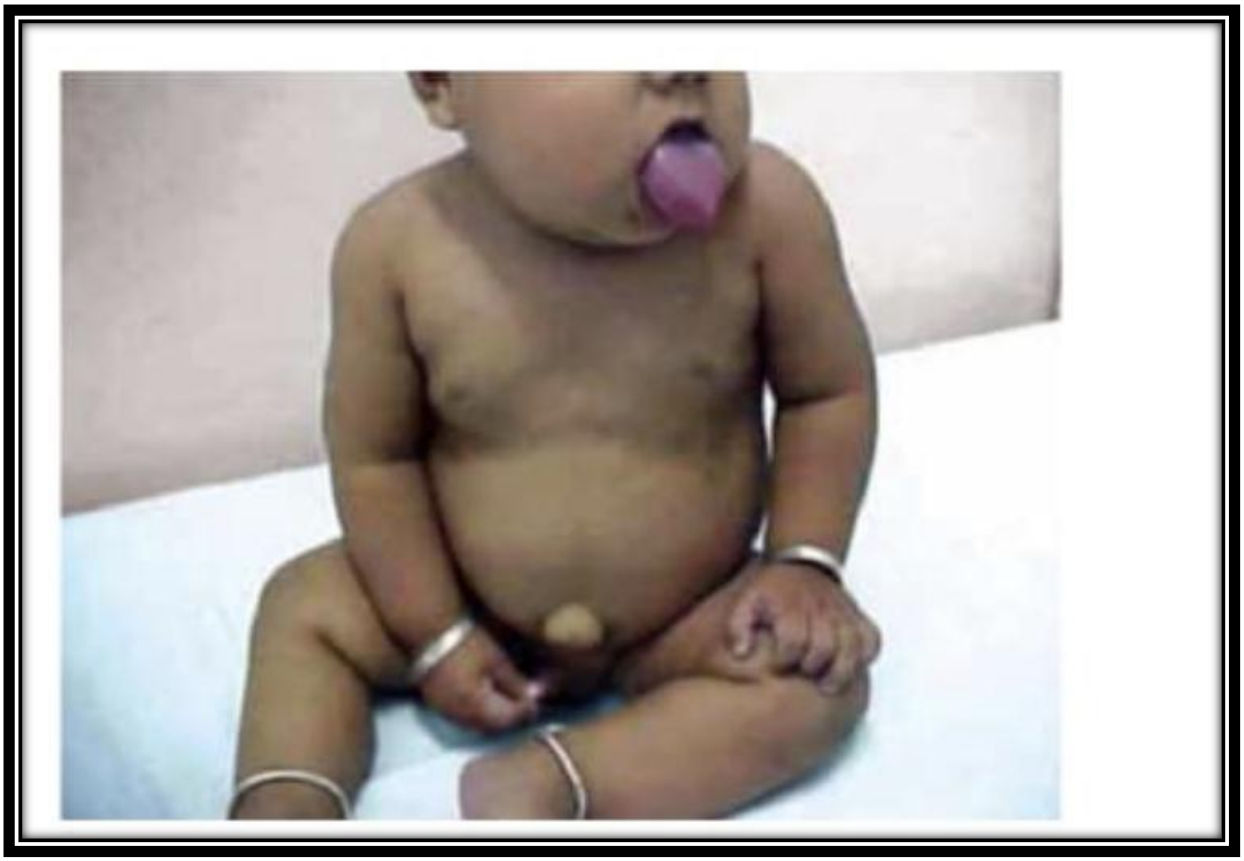


A ten month old male infant was brought with a large tongue. His birth weight was 4.2 kgs and had no perinatal or subsequent complications

1. What is the most probable diagnosis?
2. What are the components of this syndrome?
3. Which chromosome carries the gene responsible for this disorder?
4. What complications do you expect in such a child?

Answers

1. Beckwith weidmann syndrome.

2. Omphalocele, macroglossia, microcephaly, visceromegaly, hemihypertrophy.

3. 11(11p15.5).(1)

4. Tumors: Wilm''s tumor, hepatoblastoma, gonadoblastoma, adrenal carcinoma, rhabdomyosarcoma.

PROCEDURE PAGE

Bone Marrow Aspiration and Biopsy

Bone marrow examination provides crucial information in the diagnosis of various hematologic and oncologic conditions in children. Bone marrow aspiration also permits immune-phenotyping, cytogenetic analysis, and other molecular studies.

Indications:

- Pancytopenia
- Unexplained anemia, leukopenia, or thrombocytopenia (aspiration only).
- Acute or chronic leukemia (aspiration only).
- Myelodysplasia, Myeloproliferative disease.
- Non-Hodgkin or Hodgkin lymphoma.
- Childhood solid tumors (including sarcoma, Wilms tumor, neuroblastoma, germ cell tumor).
- Bone marrow failure (including acquired aplastic anemia, Fanconi anemia, Diamond-Blackfan syndrome).
- Fever of unknown origin.
- Storage disease
- Monitoring during chemotherapy or following stem cell transplantation (aspiration only).

Equipment

For Site Preparation	For Marrow Aspiration and Biopsy
10% povidone-iodine. Alcohol swabs. Sterile gloves, gown, and drape. Spinal and subcutaneous needles, 20 to 26 gauge. 1% lidocaine hydrochloride, injection. 8.4% sodium bicarbonate, injection, USP.	Sodium heparin injection Bone marrow aspiration needles (15 and 18 gauge). Bone marrow biopsy needles (11 13 gauge, 4/2 inches) Sterile syringes, 10 to 20 mL. Container with fixative for trephine biopsy specimen. sodium heparin and EDTA collecting tubes Gauze and Bandages

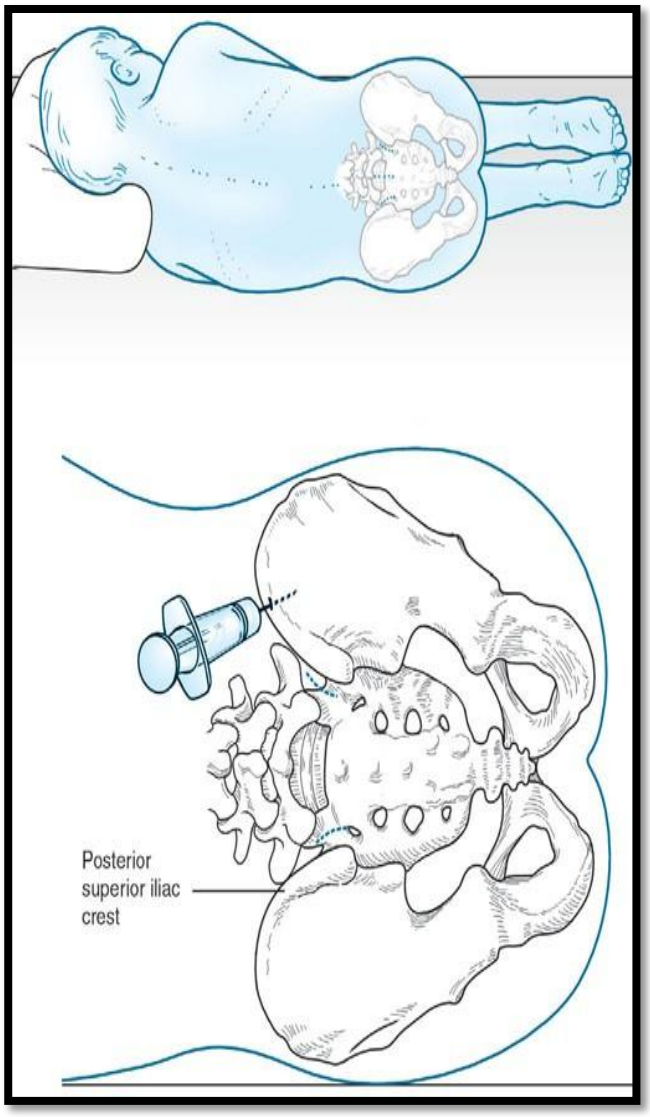
Patient preparation & Position

1. Obtain a thorough medical history and perform physical examination.

2. Obtain written informed consent.

3. Inform patients who are not receiving general anesthesia that an unpleasant, lightening-like sensation down their lower extremities may be felt at the time of suction during aspiration.

4. The posterior superior iliac crest is the optimal location for bone marrow aspiration and biopsy in most children (Fig.1)



5. The posterior superior iliac crest can usually be identified by a dimple in the skin located at the lateral edge of Michaelis' rhomboid.

6. Michaelis' rhomboid is a diamond-shaped area over the posterior aspect of the pelvis formed by the posterior superior iliac spines, the gluteal muscles, and the groove at the distal end of the vertebral column. This area can be located in most patients by palpation with the thumb, even if anatomic landmarks are not visible

7. Alternative sites include the anterior iliac crest in obese patients and the tibia in infants younger than 3 months.

8.The sternum, which is used in some adults, should be avoided in children.

9. If the posterior iliac crest is used, the patient is placed in the right or left decubitus position, with the hips flexed and the knees drawn up.

10. If the anterior iliac crest is used, the patient is placed in the supine position with the hips and knees flexed.

11. Occasionally, thin patients who do not receive general anesthesia may be placed in the prone position

Procedure

Bone Marrow Aspiration

1. Clean the site with povidone-iodine followed by alcohol swab.
2. Place sterile drape.
3. Inject lidocaine intradermally with a subcutaneous needle to produce a small wheal.
4. Use a larger bore needle to push through the skin and subcutaneous tissue and inject 2–3 mL (maximum 3 mg/kg/ dose) more lidocaine along the periosteum.
5. Hold the bone aspirate needle horizontally using the index finger near the tip of the needle for control.
6. Advance the needle through the skin, subcutaneous tissue, and the surface of the cortical bone with steady pressure and a twisting motion.An abrupt decrease in resistance occurs when the needle penetrates the cortex and enters the spongy marrow cavity.
7. Advance the needle 1 cm more before the stylet is removed.

8. Attach a 10-mL or 20-mL syringe to the end of the needle and pull the plunger back quickly to aspirate approximately 0.25 mL of bone marrow.

9. If an aspirate is not obtained, replace the stylet and advance or reposition the needle.

10. This first pull contains the marrow particles or spicules that should be used for preparing initial smears.

11. A heparinized, larger syringe (30 mL) may be used to obtain additional marrow for cytogenetic analysis, flow cytometry, and other studies

Bone marrow Biopsy

1. The trephine biopsy is the preferred method to evaluate cellularity and detect bone marrow metastasis in lymphoma and many childhood solid tumors.

2. Biopsy specimen is obtained through the same incision site.

3. Hold the biopsy needle in the same manner as the aspiration needle but angle it to sample a different area from the aspiration. Advance the needle with steady pressure to the periosteum and twist into the surface of the cortical bone.

4. Remove the obturator and push the needle through the cortex using a rotating, twisting motion

until decreased resistance is met. Advance the needle another 1–2 cm.

5. Reinsert the obturator until resistance is met to gauge the length of the specimen. Rotate the needle 360 degrees vigorously several times while moving it back and forth vertically and horizontally to break the biopsy core off the surrounding bone.

6. Carefully remove the needle and insert a separate blunt obturator into the distal end of the needle to force the core out through the hub onto a glass slide.

7. Specimen should be at least 1.5 to 2 cm in length for optimal processing.

8. If the specimen is inadequate or consists mostly of cartilage or cortical bone rather than core marrow, which appears dark red with a fine, white trabecular network, attempt additional biopsies.

9. The specimen should be placed in an appropriate fixative.

10. Apply direct pressure to the site for at least 5 minutes once the procedure is completed and the needle removed and place a pressure dressing.

Risks and complications

1. Risk of bleeding is low if adequate pressure is provided over site to achieve primary hemostasis.
2. Defects in coagulation should be corrected before the procedure.Platelet transfusion is indicated when technical difficulties are anticipated in patients, especially those who are obese, with severe thrombocytopenia.
3. Risk of infection and osteomyelitis is extremely low when procedure is performed in sterile fashion.4. Pain and discomfort are alleviated with adequate sedation and analgesics.
5. Bleeding at any site, with or without development of a hematoma, is rare if adequate pressure is applied.
6. Retroperitoneal hemorrhage, osteomyelitis, and needle breakage have also been rarely described.

Practical Points

1 Adolescents may require only local anesthesia for the procedure.
2. Conscious sedation or general anesthesia is generally necessary in young children, particularly if repeated procedures are required.
3. Adding local anesthesia in young patients also decreases postprocedural discomfort at the site.
4. Lidocaine used for local anesthesia should be buffered with sodium bicarbonate (sodium bicarbonate mixed with lidocaine in a 1:4 ratio) to reduce burning during injection.
5. Obtaining spicules (bone marrow particles rich in hematopoietic elements) on the first pull of the aspiration may be easier using a larger syringe (30 or 60 mL).
6. Aspirating more than 0.25 mL of marrow initially dilutes the sample with sinusoidal blood and interferes with morphologic studies.
7. If an aspirate is "dry" and an adequate specimen cannot be obtained, a touch imprint of the biopsy core may be helpful for cytologic examination.
8. A dry tap usually indicates myelofibrosis or a marrow cavity packed with malignant cells.

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