PBR CORRECTIONS AND CLARIFICATIONS
(Release date 10/13/12)

For the most updated corrections, clarifications and other great info, please visit the PBR forum at www.pediatricsboardreview.com/forum

Please note that these corrections and clarifications are for the first edition (called the SAR) and the second edition (now called the PBR) of the core study guide and Q&A book now found at www.pediatricsboardreview.com. For the 3rd edition of the books (release date anticipated to be in late 2012), these modifications will have already been included.

Regarding the page numbers seen below, they may be different than your page numbers because some people are referring to the online PBR, others are referring to the hardcopy PBR, others to the older online SAR and other to the oldest hardcopy SAR.

I'd just like to say thanks so much to all of you who asked such great questions. I also truly appreciate the forum posts and emails sent from docs who were just trying to help improve the PBR and made me aware of silly spelling errors or drop off sentences.

Thanks again and good luck on the exam!

- Ashish

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Page 245. The vaccine schedule reminder section and the catch up reminder section state there are only 5 vaccines given at 1 year of age.

You're correct, it's actually six. Revised version:

VACCINE SCHEDULE REMINDERS

* 2, 4 & **SIX** MONTHS: **SIX vaccines are given.** Pediarix ® (DTaP, IPV, Hepatitis B) + Hib + Pneumococcal vaccine + Rotavirus vaccine.
    - **PEARL**: The PCV-7 vaccine which is given to **all** kids. The **PPV-23 valent** pneumococcal vaccine is given to kids > 2 years of age if they have **chronic illness or asplenia**.

* 12 MONTHS: 6 vaccines are given (MMR #1, VZV #1, Hepatitis A #1, Hib #4 and the 2 boosters of PREVNAR #4 of 4, and DTaP #4 of 5).

* 4 YEARS OF AGE: Four vaccines are given around the age of 4 (MMR, Varicella, DTaP, **IPV**).
    - **PEARL**: **MMR** # 2 is scheduled around 4-6 years of age, but it **CAN** be given just **1 month** after the 1st dose! **VZV** can be given after just **3 months**!

CATCH-UP IMMUNIZATION SCHEDULE PEARLS

The CDC's catch up immunization schedule does NOT include every pediatric vaccine. For example, PCV7 and Hib are NOT recommended for kids over 5-years-old. Rotavirus vaccine is NOT given to anyone after the 12 weeks of age.

PEARL/MNEMONICS: You could be asked a question about how many **different** vaccinations/shots a child will need at a given age. You could also be asked how many shots a child will need in total for a given vaccine.

- **MNEMONIC:** SIX shots at SIX months of age and FOUR shots at FOUR years of age. For the 1 year shot, you could try to remember that SIX x 2 = 12. Therefore there are TWO major immunization milestones at which time SIX shots are given (SIX months of age and 6 x 2 = 12 months of age).
- **MNEMONIC:** 5-4-3-2-1 - Use this to help you remember the number of times various vaccinations are administered.
    - 5 = Total of 5 DTaP doses (last one at 5 years of age)
    - 4 = P = 4eumococcal & I4V = Total of 4 pneumococcal (last at 1 year of age) and IPV (last at 4 years of age) doses. A 4 also looks kind of like an H, which could remind you of Hib.
    - 3 = Total of 3 Hepatitis B doses (last at 5-6 months of age)
    - 2 = MMR, VZV, and Hepatitis A
    - 1 = Rhymes with NONE

==========

On page 47 of the hard copy, do you think it is important to know the dosages for the epi pen and epi pen jr?

Regarding the epi pens, doses are typically not tested, though epinephrine could be one of the few places you're tested on it. If you were, though, I'd think it would in more of a code type of situation. It wouldn't hurt to know it, but I'd write it in the margin and consider tying to memorize it on my 4th or 5th pass through the material.

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On page 58 of the hard copy-under IMMUNOLOGY TESTS, A RECAP, you write "an ABP fav "when testing for knowledge of agammaglobinemia. I'm confused by that statement? How does that relate to tetanus titers and B cell deficiency?

Regarding agammaglobinemia... I was talking about the fact that it's a B-cell def. So titers for tetanus and diphtheria will be low even after immunization. ABP likes for you to know that fact and can test around that information in many different ways. Here's the updated section.

TITERS

If you suspect a **B**-cell (aka Humoral) deficiency, test for it by obtaining anti**B**ody **TITERS** for something the child was already immunized against, such as **TETANUS** (testing for tetanus titers in patients with Agammaglobulinemia, a B-cell deficiency, is a an ABP favorite way of testing your knowledge). Could also test for titers against Diphtheria and Streptococcus/Pneumococcus. **Do not get confused** with getting SKIN TESTING for tetanus/Candida/Mumps/PPD which all test for T-cell mediated (aka cellular) immunity.

PEARLS:

* Infections related to B-cell deficiencies rarely occur before 6 months of age because of presence of maternal antibodies.

* ANTIBODY SUBCLASSES: If IgG levels are in the normal range but you still have a high suspicion for a B-cell/Humoral defect, consider checking for the presence of Ig subclasses for Tetanus, Pneumococcus, etc.

==========

On page 133 of the hard copy under the pearl for cardiopulmonary resuscitation (CPR) it looks like some of the writing has been left out? What comes after the but...?

Sorry about that. I must've passed out due to exhaustion :-)

CARDIOPULMONARY RESUSCITATION (CPR)

Cardiopulmonary Resuscitation (CPR) is a low-yield topic because guidelines are always changing.

* SINGLE RESCUER CPR FOR BABIES: Provide compressions and breaths at a ratio of 30:2 to minimize transition times. Also, COMPRESSIONS are more important than breaths.

* DOUBLE RESCUER CPR FOR BABIES: Provide compressions and breaths at a ratio of 15:1 (15 compressions for every breath).

* ADOLESCENTS: 30:2 regardless of the number of rescuers.

* **PEARL**: Guidelines have changed, but the key is to remember that it's becoming more and more important to focus on high quality chest compressions to get the blood flowing rather than focusing on breaths.

==========

On page 152 under section 4 (ESOPHAGEAL WEB), what are you referring to when you say the "jet phenomenon"?

When barium passes through the poorly canalized esophagus, there is a "jet" of propelled barium distal to the initial point of constriction. When looking at a barium swallow, it's the thin area of barium seen. When that area is tortuous, it can resemble a TE fistula. When it's linear, it does not. New description is below

ESOPHAGEAL WEB

An esophageal web can cause reflux-like symptoms, esophageal impaction and chest pain. It results from the failure of the esophagus to re-canalize in utero. The web then acts as an obstruction to the passage of a food bolus. Liquids, however, pass through more easily. Treatment requires dilation of the esophageal web.

IMAGES: http://bit.ly/ngX7zN and http://bit.ly/oF2ryU

PEARL: The "jet phenomenon" refers to the thin area of barium seen when looking at a barium swallow. It starts at initial point of constriction. When that area is tortuous (http://bit.ly/oF2ryU), it

can resemble a TE fistula. When it's linear it does not (http://www.ajronline.org/content/129/4/747.full.pdf - page 1 - see it and move on!).

==========

On page 91 for molluscum you say to use topical...what? is it steroids?

For once in dermatology, it's not steroids :-) Updated section below:

MOLLUSCUM CONTAGIOSUM

Molluscum contagiosum results in flesh-colored, pearly papules that are dome-shaped and **umbilicated**. Caused by the POX virus. NO treatment is needed, but sometimes cryotherapy or topical Cantharidin, Podophyllotoxin, Imiquimod or Potassium hydroxide.

==========

Please check the neonatology section under BREAST MILK. I've known for a long time that it is ok for a mom with Hepatitis B to feed her baby with precautions. You said in your book otherwise. Please check the CDC recommendations:

Is it safe for a mother infected with hepatitis B virus (HBV) to breastfeed her infant immediately after birth?
Yes. Even before the availability of hepatitis B vaccine, HBV transmission through breastfeeding was not reported. All infants born to HBV-infected mothers should receive hepatitis B immune globulin and the first dose of hepatitis B vaccine within 12 hours of birth. The second dose of vaccine should be given at aged 1–2 months, and the third dose at aged 6 months. The infant should be tested after completion of the vaccine series, at aged 9–18 months (generally at the next well-child visit), to determine if the vaccine worked and the infant is not infected with HBV through exposure to the mother's blood during the birth process. However, there is no need to delay breastfeeding until the infant is fully immunized. All mothers who breastfeed should take good care of their nipples to avoid cracking and bleeding.

Another PBR member also pointed it out and gave me an AAP reference (http://www2.aap.org/breastfeeding/policyOnBreastfeedingAndUseOfHumanMilk.html). You are both correct. The next version of the book will have this correction with Hepatitis B omitted, and also a CMV correction. Here's the most updated version:

BREAST MILK

Breast milk contains arachidonic acid, DHA, whey, casein, colostrum, hind milk, etc. It's a lot to remember, so memorize the following and move on!

* ARACHIDONIC ACID (AA) & DOCOSAHEXAENOIC ACID (DHA): Help with neurologic development. Greatest in COLOSTRUM. Not as much in mature milk.

* WHEY: The primary protein in breast MILK.

* CASEIN: The primary protein in FORMULA.

* COLOSTRUM: The milk produced at the end of pregnancy and early after delivery. Only small amounts are expressed in the first few days until the more mature milk finally comes in.

- Yellow color is from carotene.
- Stimulates passage of meconium.
- High in PROTEIN (immunoglobulins, especially IgA).

* HIND MILK: Last bit of milk expressed during breast-feeding. It is highest in CALORIES and FAT.

* FROZEN BREAST MILK: Good for 3-6 months. Once thawed, use within 48 hours.

* CONTRAINDICATIONS TO BREAST-FEEDING: Mother with herpes simplex virus (HSV), HIV, tuberculosis (TB), on chemotherapy, on HYPERthyroid medications, on metronidazole, on sulfa drugs or on Tetracycline. Breast-feeding is also usually contraindicated if the baby has an **INBORN ERROR OF METABOLISM**. An inverted nipple may be a contraindication depending on the degree of inversion. Breast shells may be needed.

PEARL: Candidiasis, mastitis and fibrocystic disease are NOT contraindications.

PEARL: Breastfeeding is NOT a contraindication for Hepatitis B. For mothers who are CMV **carriers** (not recent converters), they may also breastfeed.

MNEMONICS:

- COLOSTRUM: Although it is supplied to babies very EARLY in life, it has tremendous LONG-TERM protective benefits/ingredients (AA, DHA, IG's/IgA aka protein).
- MATURE MILK is the regular, everyday milk that provides the regular, everyday ingredients to a baby (fat, lactose, "energy," etc.).
- HIND MILK: HIND milk has a high FAT and CALORIC content, like the unusually oversized be**HIND** of an appropriately overweight/fat new mom. (Sorry for the un-PC mnemonic. Hopefully it helps).
- WHEY: A BREAST full of milk WEIGHS much more than in a can of powdered formula.

==========

In the Allergy section you stressed the use of SUBCUTANEOUS epi for food allergies as a treatment instead of IM epi.

Hi Therm,

Either can be given, but you're right. IM is better and has a faster onset of action and is more reliable. Updated section below. Thanks for the help and correction!

FOOD ALLERGIES

Early introduction of solids results in an increased chance of food allergies and may predispose children to obesity later in life. ABDOMINAL PAIN may be the only sign of impending anaphylaxis. If the patient has a history of a food allergy, GIVE IM EPINEPHRINE.

==========

In the hematology section on alpha thal, 3 missing alpha chains should be hgb H and 4 missing alpha is 4.

Thanks for the question. It's a confusing and low-yield topic so please don't spend too much time on it. You're partially correct so thank you for bringing my attention to it. The updated version is below:

ALPHA THALASSEMIA

Alpha thalassemia refers to a mutation in an alpha chain allele. Because of the defect, other types of hemoglobin persist. You will find **elevated** levels of fetal hemoglobin (hgb F) as well as elevated levels of "minor adult hemoglobin" (hgb A2). There will only be low levels of A1. Patients with alpha thalassemia do fairly well and only have a mild microcytosis.

PEARL: The only thing you probably need to know for the exam is that alpha thalassemia can cause a microcytic anemia and **cannot** be diagnosed by hemoglobin electrophoresis.

MNEMONIC: Be familiar with, but do not memorize this. It's doubtful that you will be asked to name the types of alpha thalassemia on the exam, but if you are, try this – "silent… trait (or minor)… barts… DEAD!" If one allele is mutated the patient is said to be a "silent carrier", if 2 = trait or minor, 3 = Hemoglobin H disease (or Barts) and 4 = Major (or hydrops fetalis)! Four defective alleles are not compatible with life since ALL types of hemoglobin have alpha chains. Hgb A1 = alpha-beta, Hgb S = alpha-beta (but with a defective beta due to glu-val), Hgb A2 = alpha-delta and Hgb F = alpha-gamma.

==========

I found a typo in the cardio chapter on page 69 in transposition of great arteries, the lv leads to the PA (not PV)

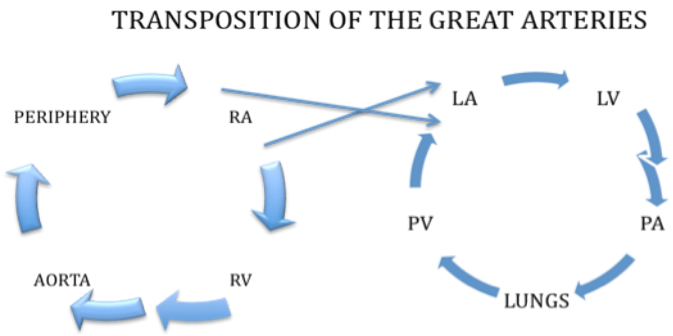
Shoot! You're right. It's correct everywhere else, including the diagram, but I did say PV at the beginning. Thanks so much for the catch… Keep em coming!

Corrected version below:

TRANSPOSITION OF THE GREAT ARTERIES (TGA/TOGA)

The "great arteries" are the AORTA and the PULMONARY ARTERY. In Transposition Of The Great Arteries, the LV leads to the PA and the RV leads to the Aorta. Most common cardiac cause for cyanosis on **DOL 1**, and usually presents **within hours**. EKG shows RVH. The two circuits do not connect and are "running in parallel" (see image). Mixing needs to occur in order to support life. Often a VSD is present, but if not, then a septal "defect" needs to be created. To treat, **create an ASD** to allow mixing. Mixing at the PDA also helps (though not as much) so create the presence of BOTH (**ASD and the PDA)** by also giving **PGE**. The ASD (or existing VSD) allows a RIGHT to LEFT shunt (deoxygenated circuit to oxygenated circuit) to be created. CXR shows an **EGG SHAPED and vascular congestion** (due to blood flow from the LV to the

PA). There is no associated murmur. In the image below, note the circuits running in parallel. Treatment is an ASD (represented by the crossed arrows).



PEARL: If you suspect a cardiac cause for cyanosis on DOL 1, this is probably your answer!

==========

In another resource, for X-linked familial hypophosphatemic rickets that the 1,25 vit D level is normal. However in your book you state that 1,25 levels are low. Is this correct?

I think your other resource is incorrect. By virtue of the mechanism, discussed in the description ("There is a defect of phosphate reabsorption in the proximal tubule AND a defect of the kidney to **convert 25-Vitamin D to** 1,25 Vitamin D ") there has to be a LOW 1,25 level.

What's the resource ☺

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In the emergency medicine section under Digoxin toxicity you state that there is an increased chance of digoxin toxicity if a patient has low K, Mg, poor renal function or O2. Does that refer to low Mg and decreased renal function and what about the O2?

Yes. I was referring to poor renal function and hypoxia. I would say just take my word for it and spend your time on more high-yield information.

Here's the updated section.

DIGOXIN TOXICITY

Digoxin toxicity leads to anorexia, weakness, n/v, poor appetite, **V-tach**. Increased chance of dig toxicity if LOW K+, Mg, poor renal function or hypoxia. Treat with BB, Digibind if life threatening correction of electrolytes.

==========

Alpers is not adrenogenital syndrome (CAH is). Alpers is progressive sclerosing poliodystrophy.

Not sure how that happened. I'm guessing lots of people noted it and said, "what's he on?"

I've made the correction. Revised mnemonic is below

AUTOSOMAL RECESSIVE MNEMONIC

This autosomal recessive mnemonic is strange, but easy to remember: "**PAT HAS WACK GAS** which made me **HURL** in the **BACK SEAT** of an **AUTO**mobile!" If you are unfamiliar with PAT, s/he is unique Saturday Night Live character. Possibly male… possibly female… definitely full of WACK GAS that would make you HURL in the BACK SEAT of an AUTOmobile!

* PAT: http://upload.wikimedia.org/wikipedia/en/9/99/Itispat.jpg
* P = PHENYLKETONURIA (aka PKU)
* A = ALPHA-1-ANTITRYPSIN DEFICIENCY
* T = TAY-SACHS DISEASE
* H & HURL = HURLER'S SYNDROME
* A = ATAXIA TELANGIECTASIA
* S = SICKLE CELL DISEASE and the THALASSEMIAS
* W = WILSONS DISEASE
* A = **ALP**ERS SYNDROME (AKA Progressive Sclerosing Poliodystrophy): This is a progressive neurologic disease. Patients do not meet their milestones and are noted to have ataxia, cognitive deficits and seizures. Also have liver disease. Die by 10 years of age.
    - **MNEMONIC**: The symptoms of ataxia and cognitive deficits are remarkably similar to how you would feel if you got ALTITUDE sickness in the Swiss **ALP**S!
* C = CYSTIC FIBROSIS
* K = KARTAGENER SYNDROME: = Immotile Celia & Sperm. Lung issues and infertile. "while playing KAR-TAG, atul's CELICA became immotile"
* G = GALACTOSEMIA
* A = see "A" disorders above
* S = see "S" disorder above
* BACK SEAT = May help you remember that this mnemonic deals with **recessive** disorders.
* AUTOmobile = AUTOsomal recessive.

==========

In the ID section under staccato, barky & paroxysmal coughs, the most common cause of croup should be viral and parainfluenza.

Thanks! Luckily the Summary Table is accurate. I've modified the two sections in the book for future members. Here it is:

(DOUBLE TAKE) STACCATO, BARKY & PAROXYSMAL COUGHS

* **STACCATO**: If this term is used to describe an infection-related cough, pick CHLAMYDIA PNEUMONIAE.

* **BARKY**: If this term is used to describe the cough, pick CROUP. Which is most commonly from parainfluenza and other viruses.

* **PAROXYSMAL**: If this term is used to describe the cough, pick PERTUSSIS.

==========

Was also looking at p. 260. The OTC explanation is not clear with PORNOthine carboxylase. I don't think there's an enzyme with that name, and I could not make sense of that sentence. I could get PORTHINE transcarbamylaX and EROTIC acid - that would be a good mnemonic.

Yup. There was a type-o, and I think the mnemonic could be stronger. I'm changing it to the following for the next print and online guide. Thanks!

ORNITHINE TRANSCARBAMYLASE DEFICIENCY

In ornithine transcarbamylase deficiency, the serum is completely void of citrulline and arginine because OTC is not catalyzing the reaction needed to make citrulline! This is the most common defect and it's X-linked. Much more symptomatic in boys than the girl carriers who only get symptoms when they are sick. Look for a **HIGH urine Orotic Acid** (all of the built up carbamoyl phosphate gets turned into this).

MNEMONICS: For **orni**thine transcarbamyla**X** deficiency**,** think **PORNO**thine transcarboxylase deficiency (or PORNO THEME transcarboxylase deficiency). Who watches EROTIC (Orotic) movies? Boys! That should remind you that it's an X-linked disorder. "Erotic" should remind you to look for an elevated OROTIC acid.

==========

I believe the current standard for Graves is no longer PTU because of the hepatotoxicity. I have been using the mnemonic Potentially Toxic Ugh.

Thanks bdig.

While it's still used, it seems to have fallen out of favor as a first-line agent. Methimazole seems to have stepped up to the plate. Love the mnemonic… I'm assuming it's yours and I'm going to use it! Here's the updated version:

GRAVES DISEASE = HYPERthyroidism

Graves disease (aka Grave's Disease) causes hyperthyroidism due to the presence of "thyroid-stimulating immunoglobulin." Signs/symptoms may include an infiltrative ophthalmopathy, emotional lability, weight loss, heat intolerance and possible LID LAG. TSH should be VERY low/absent! Radioactive Iodine Uptake is HIGH in Grave's since it needs lots of iodine to make all of the THYROXINE being released. Treatments options include methimazole, iodine ablation, a beta blocker (propranolol for symptomatic relief) and thyroidectomy. Methimazole and PTU inhibit T4 production (not secretion). GOITERS in patients with hyperthyroidism are cells that are FULL of thyroid hormone so it can take months to become euthyroid.

PEARL: Some of the symptoms of hyperthyroidism may be disguised as "hyperactivity, disorganized thinking and trouble sleeping."

PEARL: If a patient has a goiter, more information is needed to differentiate between hypothyroidism and hyperthyroidism

MNEMONIC: PTU can be quite toxic so it is NOT a first-line agent. P-T-U = Potentially Toxic, UGH!

==========

In the neonatology section, the current recs for car seat position are to keep kids rear-facing until they are 2. No longer 12 months.

Thanks! It looks like the new guidelines came out right before my first print. I've made the correction. Here's the update, and a good resource – http://www.cdc.gov/motorvehiclesafety/child_passenger_safety/cps-factsheet.html

AUTOMOBILE AND CAR SEAT SAFETY

Automobile and car seat safety guidelines vary depending on a child's age and weight.

* REAR-FACING CAR SEAT: From birth to 24 months of age, infants should be seated in the back seat at a 45-degree angle. They should be seated in the middle and face the rear of the car. This should be continued until the child is over 24 months or until they outgrow the manufacturer's weight and height limits. After that, the car seat may face forward.

* FORWARD FACING CAR SEAT: When they graduate from the rear-facing seat, they can be in a front-facing car seat with a harness until they again outgrow the manufacturer's weight and height limits (usually around 4 years of age).

* BOOSTER SEAT: Around 4 years of age or 40 lbs, kids outgrow the car seat and will need a belt-positioned booster seat. They stay in that until a regular seatbelt fits.

* SEATBELT: Children may use a regular seatbelt, while still sitting in the back seat, once it fits properly. Usually when they reach 4 feet 9 inches around the ages of 8 – 12.

* FRONT SEAT: Children may sit in the front seat once they become teenagers (regardless of weight).

==========

In adolescents, I believe the CPR ratio is 30:2 - the same ratio for everyone not a baby and not in a healthcare setting.

You're right. Thanks for the catch. Here's the updated section:

CARDIOPULMONARY RESUSCITATION (CPR)

Cardiopulmonary Resuscitation (CPR) is a low-yield topic because guidelines are always changing.

* SINGLE RESCUER CPR FOR BABIES: Provide compressions and breaths at a ratio of 30:2 to minimize transition times. Also, COMPRESSIONS are more important than breaths.

* DOUBLE RESCUER CPR FOR BABIES: Provide compressions and breaths at a ratio of 15:1 (15 compressions for every breath).

* ADOLESCENTS: 30:2 regardless of the number of rescuers.

> * **PEARL**: Guidelines have changed, but the key is to remember that it's becoming more and more important to focus on high quality chest compressions to get the blood flowing rather than focusing on breaths.

==========

Was looking over the concussion section in the ER section. Not sure how picky the ABP will be on this issue, but I believe the system you enumerated is no longer current. I don't believe that anyone is grading the concussions any longer - at least that's how we're practicing now ourselves and in conjunction with the specialists.

For this topic, it really depends on where you look. UpToDate has a VERY comprehensive discussion, but it's too difficult to simplify. The American Academy of Neurology is currently working on a new set of guidelines, but still refers physicians to their 1997 guidelines – http://www.aan.com/professionals/practice/guidelines/pda/Concussion_sports.pdf

For now, I'm fairly comfortable with the way I've written it. I think it's a good way to evaluate "ABP patients" and will likely lead you to the correct answer. Here is the section again, no changes made:

POST CONCUSSION TREATMENT

Post concussion treatment varies depending on whether there was loss of consciousness and/or amnesia:

* GRADE 1: NO LOC + NO amnesia **+ Confusion**. Reevaluate every 5 minutes for neurologic changes. May return to sports if mentation is clear and there are no symptoms for 20 minutes.

* GRADE 2: NO LOC **+ AMNESIA + Confusion**. Patient should have medical follow-up at 24 hours. May return to sports once the patient is asymptomatic for 1 week.

* GRADE 3: **LOC + AMNESIA + Confusion**. Take these patients to the ER for further evaluation and probable imaging. May return to sports once the patient is asymptomatic for 2 weeks.

  - PROGNOSIS: Worse if concussion was associated with amnesia, prolonged confusion, prolonged recovery or repeated trauma.

* IMAGING: Required if loss of consciousness (LOC) > 1 minute, or if there are still neurologic symptoms at presentation in the ER.

* **MNEMONIC**: Instead of "LOC," think "LAC" for the 3 differentiating factors (**L**OC, **A**mnesia and **C**onfusion). Grades 1 through 3 add one finding per grade, and they happen to go in the reverse order of C-A-L (Gr 1 = Confusion, Gr 2 = Confusion and Amnesia, Gr 3 = Confusion, Amnesia and LOC). Regarding timings for return to play, they are 0 weeks, 1 week and then 2 weeks of symptom-free time.

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In the rheumatology section, Oligo- and poly-articular should be the subtype names for JRA. Also, polyarticular is 5 or MORE joints.

Thanks bdig. I've rephrased to make it more clear. Also, changed the typo from LESS to MORE. Revised version is below.

JUVENILE RHEUMATOID ARTHRITIS (JRA)

KNOW JUVENILE RHEUMATOID ARTHRITIS WELL! The diagnosis requires knowing quite a few details. The child should have been under the age of 16 at the time of **symptom onset**. Symptoms must be present for at least **6 WEEKS** before the diagnosis can be made. In children, if arthritis is present it is more common in the **LARGE joints** and rheumatoid nodules are much less common when compared to adults. A positive rheumatoid factor indicates a worse **prognosis**. Do NOT order a rheumatoid factor for diagnostic purposes. It can **help with prognosis/subtyping, but a negative RF does NOT rule out RA**.

***** **O**LIG**O**ARTICULAR JRA (OLIGOARTHRITIS): This refers to JRA that affects **4 OR LESS JOINTS**, and is the more common type of JRA (>50%). Positive markers (rheumatoid factor and ANA) make this subtype much more likely. It's more common in younger girls and is associated with chronic uveitis. Since **visual complaints may be absent,** patients need to have **regular eye exams**. Boys have a better prognosis.

- **MNEMONIC**: The **O's** for **O**lig**O** look like EYES and need regular eye exams because it is the more serious subtype.

***** POLYARTICUIAR JRA (POLYARTHRITIS): This refers to JRA that affects **5 OR MORE JOINTS**. Also more common in young girls. Systemic symptoms outside of the joints is not common.

***** SYSTEMIC (aka STILLS DISEASE): This is equally as common in boys and girls. There are many classic symptoms and finding to be aware of including an episodic, salmon-colored "**EVANESCENT RASH.**" Patients may also have an **extremely high LEUKOCYTOSIS** (> 30K), with **spiking fevers, lymphadenopathy** and **possible hepatosplenomegaly**. Patients may also have **pleurisy**, **pericarditis** and the **Koebner phenomenon** (linear skin lesions appearing along a site of injury, rubbing or scratching). Serum **markers are NEGATIVE**.

- **PEARL**: **If everything else fits and the patient doesn't have an arthritis, go ahead and pick this diagnosis!** The other symptoms are commonly present well before the arthritis component kicks in.
- **PEARL**: This can be a difficult diagnosis to make and is often missed in clinical practice and on the pediatric board exam. **Please be VERY, VERY comfortable with this topic.**
- **PEARLS:** In comparison to leukemia, pain is in the AM (not at night), pain is in the joints (not the bone), mild hematologic anomalies (not severe), symptoms wax and wane (not persistent/worsening), symptoms are insidious in onset (not acute) and JRA may have a

rash. **BOTH can have lymphadenopathy and hepatosplenomegaly**. In comparison to septic arthritis, remember the insidious onset of symptoms for JRA (not acute).

* TREATMENT: First line treatment = NSAIDS. Second line treatment = STEROIDS.

==========

So, for Henoch-Schonlein Purpura I read the emedicine bibliography and discussion on HSP which seemed to waffle on the subject of steroids, though I think it's clear that steroids are used when there's GI/jt involvement:
http://emedicine.medscape.com/article/780452-medication

But, there was this article in Peds:
http://pediatrics.aappublications.org/content/early/2010/09/20/peds.2009-3348.abstract

It also suggests that it's ok to use steroids. I know in clinical practice this is what I have seen. It sounds like it's a bit of a debatable topic, and maybe you have experience that suggest otherwise, but I thought I would mention this.

Good discussion. You're absolutely right about it being debatable. I've looked at several resources and articles now, and that seems to be the running theme. In general, it's a disease that often resolves quickly and without any pharmacologic interventions.

I'd say use NSAIDs cautiously for joint pain, especially given the concerns for renal disease associated with HSP. For patients with severe symptoms requiring hospitalization, or GI symptoms that prevent them from eating, use steroids with or without other therapies (i.e., cyclophosphamide, azathioprine, plasmapheresis, IVIG, etc.). Given that it's a debatable topic, I can "almost" assure you that you would only be asked to give NSAIDS in the **absence** of renal disease or give steroids for severe symptoms.

Updated version is below:

HENOCH SCHONLEIN PURPURA (HSP)

Henoch Schonlein purpura (HSP) is a vasculitis which can involve **multiple systems**, including the skin, joints, GI tract and kidneys. Classic findings include **PALPABLE** and **TENDER** purpura that **blanch.** These are most often found at the **lower extremities and buttocks**, but may be elsewhere. There may also be **peri**articular joint involvement (soft tissue only) at the knees or ankles, and a faint rash. Patients may initially present with colicky abdominal pain +/- blood in stool +/- **intussusception +/-** gallbladder hydrops. Skin findings are impressive, but the labs show a **normal platelet count**. Urinalysis will likely show hematuria +/- proteinuria which can range from mild to nephrotic range (**order a spot protein to creatinine ratio**). This may be diagnosed on clinical findings. Complement levels can be low. A **biopsy may be obtained if there is doubt** about the diagnosis. Biopsy would show IgA, IgG and C3 deposits. For treatment, **do not give** steroid monotherapy because it will not work. The disease often resolves without intervention and the use of medications is debatable, meaning it's unlikely to be tested. For the boards, give NSAIDS for joint pain in the **absence of renal disease**, and for severe symptoms

(can't eat) or for a hospitalized patient, give steroids with or without other therapies (i.e., cyclophosphamide, azathioprine, plasmapheresis, IVIG, etc.).

IMAGE: http://bit.ly/nsNXZo

==========

I think Wegener's section at the end of rheum and right before the beginning of pulm has the hypermobility description attached to it!

You are correct sir! I've moved it to the ortho section.

New Wegener's section:

WEGENER'S GRANULOMATOSIS

(Rare in kids, low yield). Wegener's granulomatosis is a multisystem vasculitis that affects the sinuses, lungs and kidneys. Look for a +C-ANCA (as opposed to the p-ANCA seen in ulcerative colitis).

New Joint Hypermobility section:

JOINT HYPERMOBILITY

A child with joint hypermobility might present with a history of "loose joints." It sounds good, but these children are actually **more prone to getting injured with sprains**. The treatment is to encourage stretching!

MNEMONIC/PEARL: They'll talk about loose JOINTS, not someone being extremely flexible. The stretching stretches the ligaments and makes them less prone to injury as the joints are all loose and sliding all over the place.

==========

okay, so for asymptomatic pneumococcal bacteremia I have now read the Occult Bacteremia topic about not needing to treat if asymptomatic. I cannot find that anywhere at all. Emedicine discussed treating but doing so at home if the child is uncomplicated and has no focal illness. Where did you get that rec?

This page on eMedicine discusses not treating it: http://emedicine.medscape.com/article/967600-overview

"Occult bacteremia now occurs in only 1 of 200 children who present with acute fever (temperature of 39o C [102.2o F] or higher) and white blood cell counts of 15,000/µL or higher. The most likely cause of bacteremia remains S pneumoniae; when there is no evidence of toxicity, such bacteremia is generally a benign, self-limited event.

Because of the extremely low yield, blood cultures are no longer routinely warranted in children aged 3-36 months who have no obvious source of infection, and empiric treatment of occult bacteremia is no longer appropriate. Almost all cases will spontaneously resolve with a low rate of subsequent focal infection. If a child remains febrile and worsens clinically, further diagnostic

evaluation and possible empiric treatment with antibiotics pending results o cultures may be considered."

I also remember having a case like this that was discussed in great length during residency. UpToDate also discusses a watchful waiting approach for asymptomatic and incidentally found pneumococcal bacteremia. For anyone reading this… STAPH AURESUS bacteremia is **always** treated. Horrendous mortality so you have to be extra careful. Echo to rule out endocarditis, etc.

Updated occult bacteremia section:

OCCULT BACTEREMIA

Streptococcus pneumoniae (Pneumococcus) is the most common etiology of occult bacteremia (no obvious source). For Streptococcal bacteremia found incidentally, if there are NO symptoms, then you do NOT need to treat!

==========

On page 39, answer #2 is about the LGA baby born after a tough breech. You seem to have the correct answer description (you describe the answer for C), but you accidentally wrote A.

Good eye. I've modified it. Basically just changed A to C and made a small modification to the answer text.

Thanks!

==========

I was thinking that the Guillain-Barre/C. jejuni connection should be mentioned somewhere in the guide. In my review, I came across that, and though I know it, it seems like a good thing to have people know, especially since I have come across a few board-type questions that mention it.

Okay great. I've added it to the PEARLS section of GBS. Revised version is below:

GUILLAIN-BARRE SYNDROME (GBS, aka ACUTE INFLAMMATORY DEMYELINATING POLYNEUROPATHY or AIDP)

Patients suffering from Guillain-Barre syndrome (GBS, aka acute inflammatory demyelinating polyneuropathy or AIDP) may initially complain of back pain, **fever** and can have a facial palsy and proximal muscle weakness (trouble rising from chair or shrugging shoulders) prior to lower extremity symptoms. Classically, though, it is an ascending paralysis over several **days to weeks** in which there is ataxia and then an inability to walk. Look for diminished or **absent reflexes in the lower extremities** on exam. **Sensation is preserved** (as is bowel and bladder continence). It can progress to respiratory compromise requiring intubation. Perform a lumbar puncture to look for **albuminocytologic dissociation (increased CSF protein in the absence increased WBCs)**. FYI… they could say there is an absence of pleocytosis (pleocytosis means an increase in WBC's). For treatment, you can try IVIG or plasmapheresis.

PEARLS: Steroids DO NOT help. Pulse oximetry is a poor indicator of neuromuscular respiratory insufficiency. You can, however, try to obtain a negative inspiratory flow (NIF) or a Forced Vital Capacity (FVC) if the child is old enough to participate with the test (at least 5 years of age). Always keep **tick paralysis** in your differential, especially if they mention the summer time, a recent vacation, or the woods! Additionally, if someone presents with GBS a few weeks after a diarrheal illness, they might be referring to C. jejuni infection (known antecedent to GBS though mechanism is not understood). Also, when compared to any **CORD COMPRESSION SYNDROME**, GBS maintains rectal tone, bowel/bladder continence and sensation. It also has **decreased** reflexes. In cord compression syndromes, sensation, tone and continence are lost, and reflexes are **increased**.

==========

I came across some conflicting info on thyroglossal duct cysts. Here's a link to emedicine's thought on it. - http://emedicine.medscape.com/article/837477-overview#aw2aab6b4

Great catch. Here's the updated version:

THYROGLOSSAL DUCT CYST

A thyroglossal duct cyst is a midline lesion on anterior neck. As many as half of all thyroglossal duct cysts can get infected, which then increases the chances of recurrence. Therefore the treatment of choice is surgical excision. For the exam, if they describe a midline cystic lesion, choose this as the diagnosis, and **remove it**!

PEARL: Don't get confused with a RANULA: Painless mucous CYSTIC usually near the inner lips or under the tongue. Might be midline. Clear contents. Treated by removal.

==========

In the orthopedic section, the last sentence for a dislocated shoulder is incomplete (assume it's because they...)

Hmm… I can't seem to figure out what I was thinking. Sorry about that. I've deleted that fragment… Revised version:

DISLOCATED SHOULDER

If a prepubertal child presents with a dislocated shoulder, **anterior dislocation is MUCH more common than posterior**. Obtain an AXILLARY view X-ray.

PEARL: If a **prepubertal** child presents with what looks like dislocated shoulder, it's probably NOT A DISLOCATION. It's actually a FRACTURE.

==========

What's the rest of the sentence under subluxed radial head where it says "flexed and close to the body, with ."?

I think I was trying to give a more detailed description of what a child might look like in a picture. Revised version below:

SUBLUXED RADIAL HEAD (aka NURSEMAIDS ELBOW)

A subluxed radial head (aka nursemaids elbow) usually occurs in young children when a child is picked up or pulled by the arm. The forearm will be **pronated** and the arm will be **flexed and close to body**. It almost looks like the patient is wearing an invisible cast, and sometimes patient's are noted to hold the affected elbow with the unaffected hand. Treat with **forced supination**.

PEARLS: **Know** that it involves the **annular ligament** (it slips over the radial head allowing radial head dislocation). Also, it's fine to look and feel for fractures, but there is NO NEED for imaging if the story fits.

IMAGE: http://bit.ly/o3sMYW

IMAGE: http://bit.ly/p6LkXH

==========

One other thing - I have been looking at several different guidelines RE: DDH and CT after age 4 months. In practice, I have never seen a CT done. Here's the medscape article: http://emedicine.medscape.com/article/408225-overview#showall and http://emedicine.medscape.com/article/1248135-workup#a0720.

So, the wording on the DDH question in the Q&A book (#8 on p. 14 online) should be x-ray if our thoughts on CT for DDH are correct.

You'r right. I reviewed the AAP guidelines in a Pediatrics article. No mention of CT. Just radiographs. - http://pediatrics.aappublications.org/content/105/4/896.full.pdf+html

Here's the revised section from the book. I've also modified the Q&A book.

DEVELOPMENTAL DYSPLASIA OF THE HIP (DDH)

Infants with developmental dysplasia of the hip (DDH) may be noted to have a leg-length discrepancy or extra creases at the thigh. Girls and breech deliveries pose a higher risk. **Workup varies** by age and family history. Treatment requires a Pavlik harness.

* **CRITERIA FOR A CHILD LESS THAN 4 MONTHS:**
  - **ASYMPTOMATIC:** If there is a history of DDH in a **first degree relative**, get an **ultrasound**!
  - **SYMPTOMATIC:** If there are signs of developmental dysplasia of the hip (DDH) on exam, get an **ultrasound!**

* **CRITERIA FOR A CHILD GREATER THAN 4 MONTHS:**
  - **ASYMPTOMATIC**: If there is no family history, then **no workup** is needed. If there is a history of DDH in a **first degree relative** and an evaluation was never done, then since

the child is now greater than 4 months of age, **plain radiographs (hip x-rays)** would be indicated. Plain x-rays are more reliable than an ultrasound at this age (femoral head ossification centers are more developed).

- **SYMPTOMATIC:** For any child greater than 4 months of age that has signs of developmental dysplasia of the hip (DDH) on examination (regardless of family history), they need to be worked up. Therefore, **get plain radiographs** (not an ultrasound) since they're more reliable at this age.

* **PEARLS:** The age cutoff for ultrasounds is 4 months, **NOT** 6. When it comes to the Barlow and Ortolani signs, **EITHER of them being positive should prompt a workup**!

* **IMAGE**: http://bit.ly/qHTScy

* **IMAGE**: http://bit.ly/qTQrK8

==========

It says in one section that the number one etiology of CP is NOT asphyxia, but rather infection. However, the number one etiology is actually preterm/premature birth.

You're correct. The number 1 association with CP is actually prematurity. Infection is #2 or #3. Great catch. Here's the revised version:

SPASTIC CEREBRAL PALSY (CP)

Spastic cerebral palsy (CP) is a **motor impairment** due to brain lesions or anomalies. The condition that does NOT progress, but the motor component can change with time. The diagnosis is usually made by 1 year of age. **Intelligence can be fully intact**. The increased incidence is due to improved survival of preterm infants.

* SPASTIC HEMIPLEGIA: Arms are affected (good cognitive prognosis).

* SPASTIC DIPLEGIA: Legs are affected (great cognitive prognosis).

* SPASTIC QUADRIPLEGIA: All extremities are affected (horrible prognosis)

* **PEARL**: Though asphyxia is often thought to be the most common etiology for cerebral palsy, it's **NOT**. It's actually only responsible for a small number of cases. **Prematurity, IUGR and intrauterine infections** have a much higher association with cases of cerebral palsy.

==========

There is a statement in neonatology on vit D requirement for babies and kids. Can you rectify or confirm 200IU/day. I think the new recommendation says 400IU.

Thanks! You're right… Here's the AAP guideline - http://pediatrics.aappublications.org/content/122/5/1142.full

Here's the revised version:

EXCLUSIVELY BREAST FED BABIES

In exclusively breast fed babies, look for Vitamin D and Vitamin K deficiencies.

PEARL: ALL BABIES/KIDS are now supposed to get a total of 400 IU (international units) of Vitamin D per day in their diet. For formula fed babies, this is usually attainable through the formula. Breastfed babies need supplementation.

PEARL: Patients with CF (CYSTIC FIBROSIS) or RICKETS need > 1600 IU per day!

==========

Is anyone else confused about the DTaP question, #3 in the Q&A book?

I thought the book said that if the child has a seizure that they should not get the aP component but the answer states that they can get the aP component?

You are correct. There is an error. The answer choice for the following question is C (not B). So they can get one compoent of DTaP (the DT component, not the aP component). The explanation of the answer is still the same.

A 4-month old had a prolonged seizure 6 days after his 2-month well child visit. He received the standard 2-month vaccinations. He now returns for his 4-month visit. Which of the following statements is true regarding DTaP?

==========

Does the growth spurt for girl refers to accel. growth at tanner 2 or peak Ht velocity at tanner 3?

It refers to the peak height velocity. The accelerated growth phase is interesting, but from a test standpoint, you should memorize the peak height velocity.

==========

For the second mnemonic under Vitamin D & Its Evaluation, I think it should read "Which one is the ACTIVE form?" instead of 'inactive' form. Since the subsequent lines talk about the 1,25-OH Vit D. Right?

You are correct and thank you for the catch!

Here's the updated version:

VITAMIN D & ITS EVALUATION

The liver sends 25-Vitamin D to the kidneys where it gets hydroxylated to 1,25 Vitamin D (the active form). If looking for a **nutritional** deficiency, obtain a 25-Vitamin D level. **SUPPLEMENT with 1,25 Vitamin D** (the ACTIVE form). Typically, 25-Vit D is the first one you should check (especially if they ask for a screen).

MNEMONIC: Where is the Vitamin D produced that carries 2 numbers with it (1 and 25)? TWO organs = TWO kidneys = TWO numbers (1 and 25)!

MNEMONIC: Which one is the active form? Think of it this way… if you ingest a calcium containing food in its natural form, it will first go to the gut, then the liver, then the blood, then

finally the kidneys! So keeping the above mnemonic in mind, it's the Vitamin D with TWO numbers!

PEARLS: Here are some ways Vitamin D deficiency could present

* African-American (AA) breastfed child whose mom is not on Vitamin D supplementation

* African-American (AA) breastfed child whose mom is not getting enough sunlight

* Child with symptoms consistent with malabsorption

* Child with a history of epilepsy who is on anti-seizure medications

==========

I believe that Folic acid 0.4mg (or 400 mcg) is recommended to prevent neural tube defects, not 4 mg (http://www.cdc.gov/ncbddd/folicacid/recommendations.html)

I can see how that article might be confusing, but the dosage is 4 MILLIGRAMS, though there is no good data to prove it. Basically the article is saying that when a woman becomes of reproductive age, she should start taking 0.4 mg of folic acid per day (400 mcg). If a woman is trying to conceive but has NEVER had a child with a neural tube defect, the optimal dose is unclear, but most OBs will still give 4 mg (4000 mcg) and recommend that it be started before conception. For any woman who has HAD a child with a neural tube defect, they must absolutely take 4 mg (4000 mcg) starting at least 1 mo prior to conception. So basically… for a woman not trying to get pregnant, 0.4 mg (400 mcg), and for a woman trying to get pregnant, 4 mg (4000 mcg) of folic acid is recommended.

Hope that helps!

Here's the revised version:

FOLIC ACID

For any woman who is trying to conceive, 4 mg of folic acid per day is recommended to prevent neural tube defects. For any woman NOT trying to get pregnant, 0.4 mg (400 mcg) of folic acid per day is recommended.

MNEMONIC: **FOUR mg of FOUR**lic acid (not 0.4 mg)

==========

The question in about a 3 year old with macroglossia, microcephaly, and umbilical hernia says. Answer is Beckwith-Wiedemann Syndrome. Why can't it be hypothyroidism?

I was thinking it can have all the 3 features described in the question and was confused……

It's the microcephaly that differentiates hypothyroidsm from Beckwith-Wiedemann Syndrome. It's not a commonly associated finding in hypothyroid kids In fact, a AAP Pediatrics article even states that hypothyroid kids may have MACROcephaly compared to body size: http://pediatrics.aappublications.org/content/59/4/628.abstract

Hope that clarifies.

==========

Is the description of the dawn phenomenon correct? I have a resource that says hyper glucermia due to gh and cortisol.

Do you mean hyperglycemia? Regardless, as much as I can… I "guarantee" they will not ask you about the pathophysiology of the Dawn Phenomenon in patients with diabetes. The would test you on the treatment of the Dawn Phenomenon -> Treat by giving the nighttime insulin dose LATER than usual.

==========

Acid base section:

Posthypocapnea causes non gap metabolic acidosis and posthypercapnea causes metabolic alkalosis due to serum bicarb decrease and increase by renal compensation during prolonged resp alkalosis and acidosis respectively. This happens usually over a period of few days after correction of hypo/hypercapnea before kidneys go back to the normal reabsorption.

Also in another section, decrease in bicarb could be 2 per 10 CO2 decrease or 5 per 10 of CO2 decrease….(study guide says increase)

Ahh... you're right, and THANK YOU for bringing it to our attention.. For the first one it looks like I fell into the terminology trap, which can get confusing. For the second one, it was a copy/paste error from topic above it.
Here goes....
HypOcapnea means there is low CO2 in the blood, and this occurs due to HypERventilation.
Therefore you get a RESP ALKALOSIS and a compensatory nongap metabolic ACIDOSIS.
HypERcapnea means there is high CO2 in the blood, and this occurs due to hypOventilation.
Therefore you get a RESP ACIDOSIS and a compensatory metabolic ALKALOSIS.
Revised versions of both topics are below

NON-ANION GAP METABOLIC ACIDOSIS

Non-anion gap metabolic acidosis conditions include Ureterostomy, Small bowel fistula, Extra chloride, **DIARRHEA** (most common cause), **Carbonic anhydrase inhibitors (acetazolamide), Adrenal insufficiency**, **Renal Tubular Acidosis**, and Parenteral nutrition/Pancreatic fistula/PosthypOcapnea. Look for **HYPERCHLOREMIA** and a LOW BICARB. Here are a few mechanisms to keep in mind:

* Extra chloride: Too much saline causes a hyperchloremic non-gap metabolic acidosis

* Diarrhea: Acidosis occurs from bicarbonate loss.

* RTA: Mechanism varies.

* Carbonic Anhydrase Inhibitor: Promotes renal bicarbonate loss.

PEARL: -CAPNEA refers to how much CO2 is in the blood and is NOT referring to how fast someone is breathing. So, hyp**O**capnea means there is **low CO2** in the blood, and this occurs due to **HypERventilation**. Therefore you get a RESPIRATORY ALKALOSIS and a compensatory nongap metabolic ACIDOSIS. Hyp**ER**capnea means there is **high CO2** in the blood, and this occurs due to **hypOventilation**. Therefore you get a RESPIRATORY ACIDOSIS and a compensatory metabolic ALKALOSIS. Make sure you look carefully at the terminology in the question stem or vignette.

MNEMONIC: “Acid-azolamide”

MNEMONIC: NON-GAP CRAP! Several stool related issues cause a non-gap acidosis (diarrhea, small bowel fistula, ostomies).

MNEMONIC: USED CARP (Ureterostomy, Small bowel fistula, Extra chloride, **DIARRHEA, Acetazolamide/Adrenal insufficiency**, **Renal Tubular Acidosis**, and Parenteral nutrition/Pancreatic fistula/Poshypercapnea.

MNEMONIC: If you do not know your normal values for routine chemistries, a non-gap acidosis can be tricky. If you just calculate the gap and see that it’s normal, you could get in trouble. You should look closer at the chloride and bicarbonate levels. So, what I’m saying is that the chemistry looks good on the outside (i.e. gap looks good), but it’s actually falling apart on the inside (low bicarb and high chloride), LIKE A **USED CARp**!

MNEMONIC: Building on the one above mnemonic, a USED CARp can only travel on smooth roads WITHOUT GAPS/potholes!

===

RESPIRATORY ALKALOSIS

A respiratory alkalosis can be caused by basically anything that causes tachypnea. Sometimes this is due to hypoxia. The list includes, but is not limited to, early asthma, pneumonia, one of the aspirin phases, high altitude, fever and anxiety/hyperventilation.

PEARL: If given an ABG and you note a respiratory alkalosis, ALWAYS calculate for compensation. You do this by looking at the decrease in PCO2 from the baseline of 40, and checking to see if the compensatory decrease in bicarbonate (compensatory metabolic acidosis via bicarb excretion) is appropriate. If the bicarbonate level is even lower, then you have an additional primary metabolic acidosis. If it’s higher than expected, then there is an additional primary metabolic alkalosis. Keep in mind that the compensatory decrease in bicarbonate could be 2 per CO2 decrease of 10, or 5 per CO2 decrease of 10. That depends on whether or not you are dealing with an acute or a chronic respiratory alkalosis.

==========