Star health

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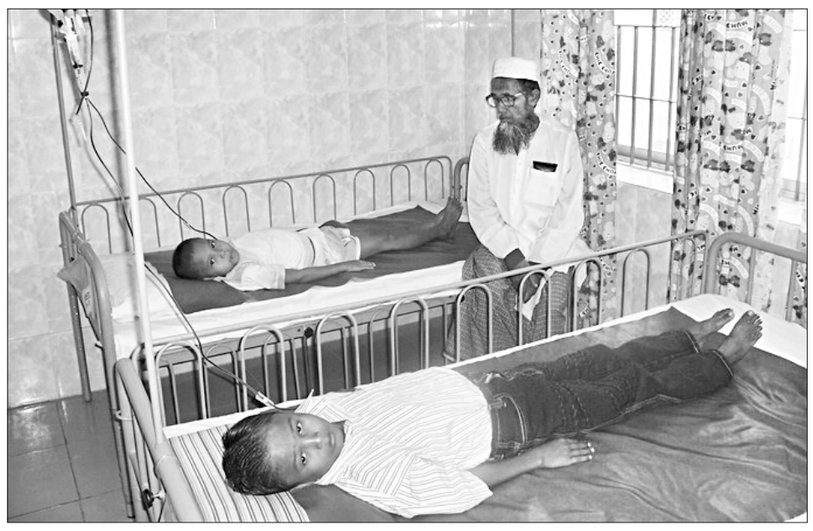
Prevention is the only way to combat thalassaemia

PROF WAQAR AHMED KHAN

Thalassaemia is the most common genetic disorder in the world and varies in different population group in the world.

A world health organisation (WHO) report estimates that 3 percent are carriers of beta thalassaemia and 4 percent are carriers of Hb E in Bangladesh. In Bangladesh more than 7000 children are born with thalassaemia each year.

A study carried out by Dhaka Shishu Hospital Thalassemia Center in 2004 in school children of Bangladesh showed that carrier status is higher and there is also regional variation. This study revealed that the overall prevalence of beta thalassemia trait in Bangladesh was 4.1% and Hb E trait 6.3%.



Children taking blood transfusion at Dhaka Shishu Hospital.

What is thalassaemia?

Thalassaemia is an inherited disorder of the blood, which is passed from parents to children. Thalassaemic patients cannot make adequate haemoglobin in their body and consequently suffer from anaemia.

There are two types of thalassaemia – (i) Alpha thalassaemia and (ii) Beta thalassaemia.

Alpha thalassemia is rarely seen in our country while beta thalassaemia is common.

Beta thalassaemia can be classified into three types – (i) Thalassaemia major, (ii) Thalassaemia intermedia and (iii) Thalassaemia minor.

Who is a carrier?

A person with one normal gene and one thalassaemia gene are said to have thalassaemia trait or to be thalassaemic carrier. A person with one normal gene and one gene for haemoglobin E are said to be Hb E trait or carrier.

It is important to identify the carriers because they themselves are healthy and lead a normal life, but may be responsible for passing serious inherited haemoglobin disorder to their children. When by chance a couple are both carrier of beta thalassaemia there is 25% chance in each pregnancy that the child will inherit the thalassemic gene from both parents resulting in a child with thalassemia and 50% chance that the child will be a healthy carrier and usually symptoms free and in 25% the child will not be a carrier and will be healthy. These probabilities exist for each child independently of what happened with prior children the couple may have had. In other words each new child has one in four chance of having thalassaemia. A couple possessing thalassemia traits can have children none of whom have two thalassemia genes. Another similar couple can have two or more children with severe thalassemia. This creates problem in genetic counseling.

Treatment

Blood transfusion: The mainstay of treatment is regular safe blood transfusion to maintain a haemoglobin level of 9-10 gm/dl and to remove the iron from the body which accumulates in the body. The iron needs to be removed as it accumulates in the liver, heart and endocrine organs ultimately leading to their failure and death.

Drugs: Two drugs which are currently in use are deferoxamine and deferiprone.

These are iron chelating drugs. Deposition of iron in the heart is the main cause of death in thalassemia. Recent trials have shown better result in removing iron from the body if combined therapy of both Desferal and Deferiprone are given.

Bone marrow transplantation: Bone marrow transplantation cures the disease but it is very costly. Non-availability of properly matched donors limits its use. Moreover the chance of graft rejection make patients confused.

Prevention of thalassaemia

In the coming years it is essential that serious efforts should be made to control thalassaemia problem and it has to be recognised as an important health issue.

Creating awareness: Awareness about the disease can be made by holding seminars, workshops, and through the mass media. 8th May is being observed as International Thalassemia Day. In Bangladesh it is also being observed by different organisations.

Population screening: Population screening to find out the carriers is an important pre-requisite for preventing the births of thalassaemic children. In many countries screening is performed and there are educational programmes in high school which is very effective.

Screening can also be done of married couples of either male or female. If one of them is a carrier then his/her partner must be tested and if positive they should be sent for genetic counseling. There must be facilities for pre natal (before delivery) diagnosis.

Genetic counseling: As part of prevention of the births of thalassaemic patients, genetic counseling plays an important role although the response may vary depending on the availability of prenatal diagnosis.

Prenatal diagnosis: Prenatal diagnosis allows a couple to abort a foetus suffering from thalassaemia major. It is usually done in the 9th to 10 th week of pregnancy. The procedure has become relatively simple. In Cyprus the number of new births of thalassaemic patients has almost fallen to zero where 80% prevention is due to prenatal diagnosis and selective abortion.

Thalassaemia situation in Bangladesh

Thalassemia patients care in Bangladesh is very poor. Ninety percent of thalassemia patients cannot afford adequate treatment. Majority of the patients cannot afford to buy the drugs. Furthermore drugs are available in the market irregularly with fluctuating cost. Besides, all drugs are not available in the market.

The government should pay attention for prevention, care and management of thalassaemic patients. Every major hospitals should have a thalassaemia center. The drugs should be subsidised. Carrier screening and prevention of births of thalassaemic children should be now a priority as it has been established that prevention of births of thalassaemic children is more cost effective than treating them. DNA lab must immediately be established in this regard.

Prof Waqar Ahmed Khan is a Professor of Pathology of Dhaka Shishu Hospital, Bangladesh Institute of Child Health and President, Dhaka Shishu Hospital Thalassemia Center.

Vitamin D lowers cancer risk

Cancer researchers urged people to take more vitamin D to lower their risk of colon, breast and ovarian cancer, saying studies showed a clear link.

"Our suggestion is for people to increase their intake," through diet or a vitamin supplement, Dr. Cedric Garland said.

Garland's research team reviewed 63 studies, including several large long-term ones, on the relationship between vitamin D and certain types of cancer worldwide between 1966 and 2004.

He said the benefit of vitamin D was as clear as the harmful link between smoking and lung cancer. "There's nothing that has this ability to prevent cancer," he said, urging governments and public health officials to do more to fortify foods with vitamin D.

The paper concluded that vitamin D deficiency may account for several thousand premature deaths from colon, breast, ovarian and other cancers annually.

Vitamin D is found in milk, as well as in some fortified orange juice, yogurt and cheeses, usually at around 100 international units (IU) a serving.

People might want to consider a vitamin supplement to raise their intake to 1000 IUs per day, Garland said, adding that it was well within the safety guidelines.

The authors said that taking more vitamin D could be especially important for people living in northern areas, which receive less vitamin D from sunshine.

SOURCE: American Journal of Public Health

Rotavirus diarrhoea: A common problem in winter

DR M KARIM KHAN

Diarrhoea is a very common problem in our country which claims so many lives each day. Rotavirus is the commonest organism causing diarrhoea all over the world. Rotavirus mostly affects the children.

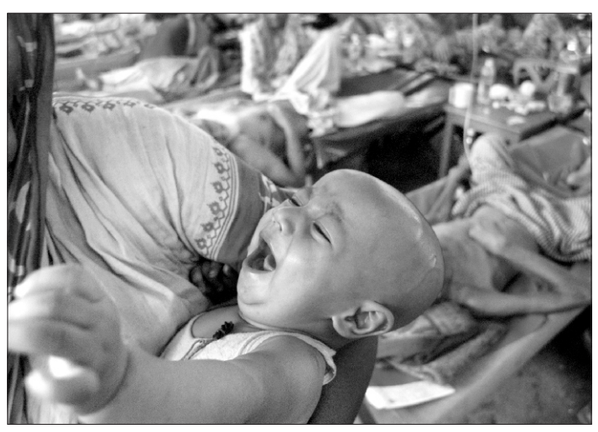
Rotavirus diarrhoea claims 600,000 deaths each year all over the world. We do not have exact statistics about the magnitude of the disease in our country, but the morbidity and mortality due to rotavirus is alarming.

Rotavirus diarrhoea mostly found in the month of November to April, which means it affects mostly in winter season.

Primary mode of transmission is faeco-oral route (). As the virus is stable in the environment, transmission can occur through infestation of contaminated water, food and contaminated surfaces.

The incubation period of rotavirus diarrhoea is approximately 2 days. The disease is characterised by vomiting and watery diarrhoea with some fever and may have some tummy pain. The duration of the episode persists for 3-8 days. Dehydration and electrolyte imbalance occurs very quickly. If rehydration measure is not taken promptly and properly there may be cessation of urine formation which may lead to acute renal failure. Immunity after infection is incomplete but repeated infection tends to be less severe then the previous infection.

Laboratory diagnosis may be done by rapid antigen detection of rotavirus in the stool specimen, but it is not practised usually. We diagnose clinically and by routine examination of stool. If there are less than 8 fecal leukocytes per high power field, we consider it as viral diarrhea.



Aim of treatment is to prevent dehydration. So oral rehydration saline (ORS) therapy to be started as soon as possible. The dosages of ORS is 5 ml per kg body weight per stool. Suppose, if a baby weighs 10 kg, having diarrhoea then, 5ml x 10 = 50 ml ORS to be given for passage of each liquid stool and to be continued till the diarrhoea is controlled.

If there is severe dehydration (urine output decreases, patient is restless or drowsy, unable to drink even, skin elasticity decreases) immediate hospitalisation is badly needed for proper management.

Prevention can be done by frequent hand washing with soap and clean water and maintaining personal hygiene.

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CONGENITAL HEART DISEASE

Be aware if your baby turns blue

DR MD HABIBE MILLAT

Children can turn blue or be blue for many reasons including heart disease. Not all children who turn blue have a heart problem.

Unfortunately, children can be born with a variety of heart defects. Generally, when people refer to "holes in the heart", they are talking about holes in the atrial or ventricular septum. However, "blue babies" rarely have just a septal problem. Often, there are other serious problems with the heart valves as well as the major vessels of the heart, lungs or other breathing problems, being cold, or having seizures.

In normal child, oxygen-rich blood travel through the lungs appears bright red. On the other hand, oxygen-poor blood appears more purplish. These children are called "blue babies" because they actually appear blue. These infants have a characteristic blue hue due to oxygen-poor blood contained in the vessels appears blue through the skin. In general, defects that impair the normal flow of blood to the lungs can result in a "blue baby".

Blueness or cyanosis can be present all the time with certain un-repaired heart problems. However, children can be pink most of the time but have "blue spells." This message concerns children who are blue all the time.

Many children with blue or cyanotic heart disease do not have the same level of energy as other children do. They require more rest. Young children tend to be very good at limiting their own activity level and resting when they need to. For this reason it is not usually necessary to force rest periods, breaks or nap times. It also takes a bit more energy to grow when a child is blue. Many of these children are on high calorie formulas. This gives them extra calories in every ounce they drink. You should consult a physician and cardiologist who can measure your child's growth and development carefully. Blue children also require more iron in their diet and are sometimes on iron drops in addition to iron-rich formula. This helps them carry oxygen in the blood more efficiently.

It is important to know your child's usual colouring and how he or she behaves on a normal day. You will know your child better than anyone. If you notice a persistent change in colour or behaviour such as increased sleeping, poor feeding, decreased energy, squatting or other changes, notify a physician. A cold that causes a lot of congestion may affect their breathing and colour. It is important to keep in contact with the physician, who will determine if any medication or other help is necessary.

Most children with blue or cyanotic heart disease do not need to use oxygen at home. Children with long lasting respiratory infections or lung problems may have oxygen prescribed for use at home. It is common for children with blue heart disease to be on iron supplements and for some to be on aspirin. Many of these children require medications for congestive heart failure as well. Fortunately, many of these heart defects, from both the cyanotic and the acyanotic categories, can be surgically repaired, thus restoring normal circulatory function.

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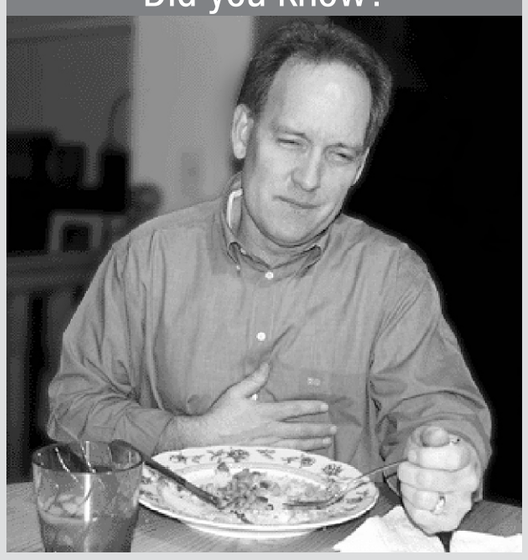
Did you know?



Later bedtime after meal may ease heartburn

A shorter dinner-to-bed interval is significantly associated with an increased risk of gastro-esophageal reflux disease, or GERD, according to researchers in Japan.

"It is generally recommended that patients with GERD refrain from eating within three hours of going to sleep," Dr. Yasuhiro Fujiwara and colleagues from Osaka City University noted in the journal.

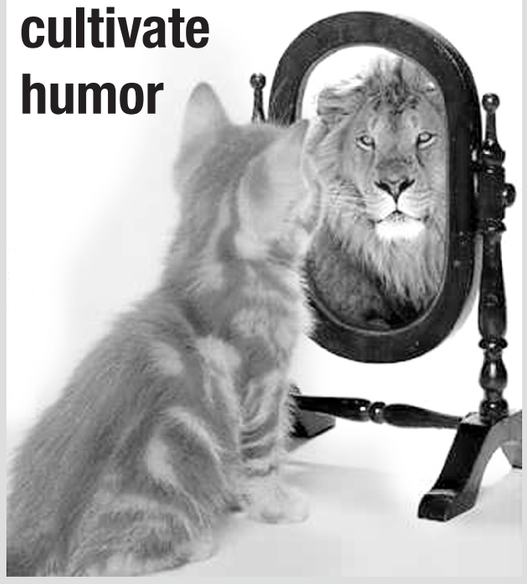
To investigate, the researchers used a questionnaire to assess the time between finishing dinner and going to bed among 147 GERD patients and 294 matched "controls" without GERD symptoms during the previous year.

A significant association was seen between shorter dinner-to-bed time and an increased risk of GERD. Participants who went to be bed within three hours after eating were 7.45 times more likely to suffer from heartburn as those whose dinner-to-bed time was four hours or longer.

SOURCE: American Journal of Gastroenterology

LAUGHTER IS THE BEST MEDICINE

Tips on how to cultivate humor



STAR HEALTH DESK

Humor does not typically come to mind in the same breath as depression. But humor can be an important ally in getting beyond the rigidity of thinking that accompanies depression and keeps people locked into a depressed state of mind.

One goal of cognitive therapy is to change the perspective and the point of view. Humor is one way to change one's view viscerally and enjoyably.

Cultivating a humorous mindset helps you see yourself and any situation with a more supple mind so that you are not locked into a negative view. Depression is both caused by and causes the inability to see options and choices we otherwise would.

Humor fosters acceptance of our humanness and our foibles. It is not sarcasm or put-downs. What we are looking for is gentle, playful perspective that embraces humanness but never at the expense of others or of ourselves. The goal is not to take life too seriously.

How to foster good humor?

- Choose to allow yourself to laugh at your own behaviors and beliefs, but not at yourself. Make that distinction clearly.

See your life not as a distraught drama but as a romantic comedy. Recognise the inherent farce-like quality in situations including sex and relationships.

Cultivating humor not only makes life more bearable, it makes you more attractive to others. Study upon study shows that a sense of humor is high up on the list of traits that most people seek in a partner.

- **Insert silliness:** Fill your life with one goofy thing a day. Make an unusual observation about someone. Or do something you normally would not do. Wear something silly. You will learn that nothing terrible happens and you may also discover that something good often happens.
- Puncture a rigid mindset with a mental exercise called "paradoxical intention."

Suppose you have to give a speech and you are unduly anxious about looking uncomfortable. You can overcome the fear of failure by deliberately focusing on it and humorously exaggerating the very effects you fear.

- Exaggeration is funny because it skewers the falsehood. If you fail at a test or perform poorly at an audition, you could erroneously call yourself a failure. That, however, is an overgeneralisation. Alternatively, you could see yourself as someone who failed at this particular thing, but in no way does that stamp you forever in this way.

Find the humor by saying, this makes me an utter wretch, a failure now and forever, a doomed and worthless subhuman, because I did not get the part that I wanted or my partner is not giving me the attention I want. Get into the exaggeration until you see the absurdity of seeing yourself as a "total failure."

- Walk down the street remembering that people are nude under their clothes. It reduces fear of others. Such thoughts can take people of high status from deity to human. It helps to remember that everyone yells at their kids, spills ketchup, goes to the bathroom.
- **Play to an audience:** Think of stories and items that would make others laugh.
- Be sensitive to the words you use. They can rigidify or help loosen up your thinking.
- Create cute, funny neologisms with your partner. Call it goofifying. Creating your own funny expressions for your experiences makes you more flexible and allows you to interpret and assess reality better.