# Ibuprofen-induced DRESS Syndrome in a Child

Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS) is a rare drug-induced hypersensitivity syndrome with life-threatening complications. Although there are a few reported cases of ibuprofen-induced DRESS syndrome in adults [1], It has not been reported in children. We report here a child with DRESS syndrome triggered by ibuprofen.

An 11-year-old boy was admitted with complaints of fever, and rash on the face and body for last five days. He had used ibuprofen for a few days due to myalgia and fever about a month ago. He was febrile and had maculopapular rash on face, body and legs. Laboratory findings were: hemoglobin 13.4 g/dL, white cell count 10400/$mm^3$, platelet count 360000/$mm^3$, absolute eosinophil count 1560/$mm^3$, total bilirubin 3.2 mg/dL, direct bilirubin 1.7 mg/dL. ESR 49 mm/h, CRP 33 mg/dL, ALT 69 IU/L, AST 63 IU/L and GGT 101 IU/L. Blood culture and throat culture were negative. Serological tests for Epstein-Barr, cytomegalovirus, HHV-6, hepatitis A, hepatitis B, hepatitis C, HIV, and parvovirus B19 were negative. Hepatobiliary ultrasonography showed normal findings. His skin rash and fever regressed, and transaminase level decreased in 48 hours after starting oral methylprednisolone (2 mg/kg/day). Three weeks later, he had no complaints, and laboratory findings were all in normal ranges.

DRESS syndrome usually occurs 2-6 weeks after exposure to the causative drug. The skin, liver, and hematological system are most commonly involved. Hematological abnormalities are leukocytosis, eosinophilia and atypical lymphocytes [2]. Other manifestations are lymph node enlargement, pneumonia, nephritis, myocarditis, encephalitis, and rarely pancreatitis. The child's score for defining DRESS syndrome was seven, and this pointed to 'definite case' [2]. Rapid response to corticosteroids also supported our diagnosis.

Differential diagnoses of DRESS syndrome include Steven-Johnson syndrome, toxic epidermal necrolysis, rheumatological diseases, and infectious diseases. Pathogenesis of DRESS syndrome is not well known. Hypersensitivity reaction secondary to circulating antibodies or toxic metabolites is implicated; herpesvirus-6 is also postulated to play a role in its etiology [3]. The incidence is approximately 1 in 1,000 to 1 in 10,000 exposures [4]. DRESS syndrome has a mortality rate of 10–20%, with most fatalities resulting from liver failure.

Drugs mostly related with DRESS are anticonvulsants, sulfa derivatives, antimicrobials and anti-inflammatory drugs, with only a few adult cases related to ibuprofen [2,5]. To our knowledge, this is probably the first report on DRESS syndrome triggered by ibuprofen in the pediatric age group.

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# Tragic Outcome of Peanut Allergy

Peanut and tree nut allergies are responsible for 80% out of 100-200 lethal cases of food allergy [1]. Children with food allergies have a lower quality of life caused by fear of a possible anaphylaxis [2,3].

We report a case of 9-year-old girl with positive family history of atopic diseases. Her first acute allergic reaction with dyspnea and vomiting occurred at the age of two years. At that time, she was hospitalized, treated with intravenous steroids, and was diagnosed with allergies to cow's milk, chicken egg and peanuts. Due to food allergies and frequent respiratory tract infections, her parents decided not to send her to a kindergarten. When the girl was 5-year-old, allergy tests did not confirm allergies to cow's milk and chicken egg anymore; however, specific IgE against peanuts were still present in high titers. One year later, she started attending an organized pre-school learning. She used to eat home-made meals only. At the age of nine years, she participated in the school camp – her first fully independent trip. She was equipped with an adrenaline auto-injector. The girl was educated and aware of her illness, and she avoided consuming peanuts. Despite that, once at home, she ate three pieces of chocolate labelled with a warning 'may contain peanuts'. After a few minutes, she developed stomach ache and dyspnea. Her father immediately administered her 0.15 mg of adrenaline intramuscularly, but she lost consciousness. Her neighbour, who was a paramedic, administered another dose of 0.15 mg of adrenalin from auto-injector and started resuscitation. The ambulance and emergency helicopter arrived within a few minutes. The child was intubated, chest compressions were carried out, and adrenaline, hydrocortisone and calcium chloride were administered intravenously. The girl regained consciousness for a short period of time. However, while she was being transported to the hospital, she again went into a cardiac arrest and despite 2.5 hour long resuscitation, she died. The postmortem report suggested anaphylaxis as the cause of her death.

Despite continuous improvement in diagnostic methods, the most important factors for patients with food allergies are more legible information on food packaging, strict diet and proximity of professional medical help. It is essential to educate patients about their allergy and equip them with adrenaline auto-injector. 3E (education, elimination, epinephrine) should be the first line of defense from a tragic results of anaphylaxis.

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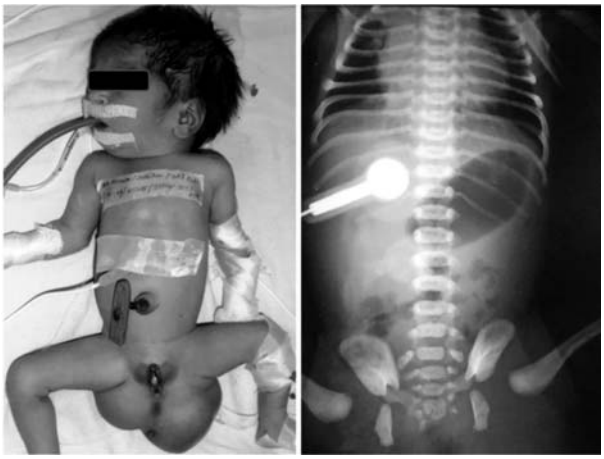
# VACTERL Association with Sacrococcygeal Teratoma

VACTERL association refers to the non-random co-occurrence (1 in 10,000 to 1 in 40,000 live-born infants) of vertebral anomalies (V), anal atresia (A), congenital heart defects (C), tracheoesophageal fistula (TE), renal anomalies (R) and limb defects (L) [1,2]. It is typically defined by the presence of at least three of them without clear evidence for an alternate, overlapping diagnosis [1-3]. The occurrence of VACTERL association with sacrococcygeal teratoma (SCT) is extremely rare.

A 2-day-old female neonate presented with a large sacrococcygeal lump with absent normal anal opening (***Fig.* 1**). The patient was in respiratory distress with nasal flaring. Examination revealed vestibular fistula, fine crepitations with presence of cardiac murmur. Failure to negotiate a red rubber catheter down the esophagus suggested the presence of esophageal atresia. Radiological evaluation confirmed the presence of esophageal

atresia with tracheoesophageal fistula (TEF), increased cardio-thoracic ratio, Altman's type I SCT (***Fig.* 1**), and left multicystic kidney. Thoracotomy with staged repair of Vogt type3b was performed. Postoperatively patient developed sclerema and died. Echocardiography to confirm the presence of cardiac anomalies, and tumor markers for teratoma were not possible due to resource constraints. In addition to SCT, we made a diagnosis of VACTERL association owing to presence of three anomalies in our patient.



*(a)* *(b)*

**FIG. 1** *(a) Neonate with sacrococcygeal teratoma type 1,small perineum and vestibular fistula, with red rubber catheter not going beyond 10 cms into the esophagus; (b) radiograph showing dilated stomach shadow, soft tissue shadow in the sacrococcygeal region, normal vertebrae and increased cardiothoracic ratio.*

VACTERL association specifically refers to the structural abnormalities derivative of the embryonic mesoderm (disruption in the proliferation, migration and differentiation of mesoderm) [1]. Epiblasts cells migrating from primitive node and proximal part of primitive streak lead to the formation of notochord, paraxial and intermediate plate mesoderm [4]. Failure of some of these epiblasts cells to migrate will lead to remnants at primitive streak which may persist in sacrococcygeal region as a teratoma[4].

We propose that VACTERL association and SCT may be more than a chance association in our patient. Clinicians should have high index of suspicion for VACTERL association in a neonate presenting with sacrococcygeal teratoma and anorectal malformation.

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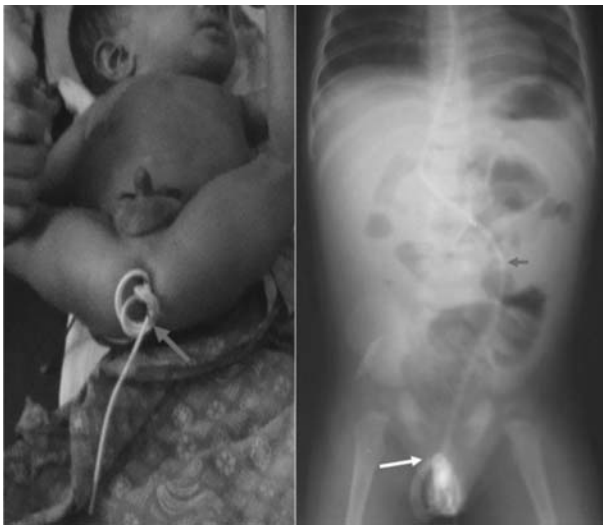
# *En-masse* Protrusion of Ventriculo-peritoneal Shunt Tube Through the Anus

A 7-month-old boy, with a right-sided Ventriculo-peritoneal (VP) shunt *in-situ* for 2 months, presented with shunt tube protruding through anus for 2 hours. The infant was treated for acute diarrhea till 2 days ago. There were no signs of meningitis or peritonitis. Perineum showed a 'bunch of shunt coils' dripping cerebrospinal fluid (***Fig.* 1**). Abdominal *X*-ray showed point of entry of the shunt tube into the sigmoid colon with no pneumoperitoneum (***Fig* 1**). The shunt was divided through small subcostal incision; cranial end was removed and the peritoneal end was pulled out through anal opening.

Besides infection, malfunction, and CSF loculations, the shunt tube can migrate into any visceral organ [1]. Intestinal perforation caused by shunt procedures is rare, and about 50% occur in infants. Anal protrusion of shunt is an extremely rare complication [1,2].

Often, some surgeons keep sufficient length of shunt tube to accommodate for the linear growth of the baby by coiling the peritoneal end and securing the 'bunch of coils' with an absorbable suture in the supra-hepatic space so that shunt does not spread itself all over the peritoneal cavity between the intestinal loops. This decreases the chances of intestinal perforations and spontaneous knotting. Despite this effective technical



*(a)* *(b)*

***Fig.* 1** *(a) Infant with 'bunch of coils' of shunt tube protruding through the anus with securing sutures in situ (red arrow); (b) Abdominal X-ray showing point of shunt entry into the sigmoid colon (red arrow) and shunt coils lying in the perianal region(white arrow).*

modification to tackle peritoneal complications, anal protrusion still occurred in this child, and the entire 'bunch of intact coils' of shunt protruded *en-masse* through the anus without any peritonitis. Such protrusion should create a big rent in the eroded sigmoid colon or cause peritonitis but strangely there was none, suggesting that shunt erosion is a slow process where erosion and healing by shunt induced adhesions takes place simultaneously to conceal a free perforation. The shunt tip adheres and erodes the bowel by continuous friction, and is then propelled distally by peristalsis to protrude anally [3,4]; diarrhea may further aggravate the process of protrusion.

Mechanisms responsible for silent erosion and anal protrusion are multifactorial. Predisposing factors for anal protrusions are stiff shunt tube, thin bowel wall with strong peristalsis in infants, malnutrition, infection and foreign body reaction. Exaggerated peristalsis in diarrhea can predispose to *en-masse* protrusion of shunt coils. Redundant sigmoid colon is the most favorable site for shunt erosion and subsequent anal protrusion. Abdominal *X*-ray does not show pneumoperitoneum because shunt perforations are usually concealed. Anal protrusion of shunt without peritonitis is treated by percutaneous division and removal of the cranial end, and the peritoneal end is pulled out through the anus [3,5]. The perforation is usually sealed by a chronic fibrous sheath around the shunt track, and laparotomy is usually not required [3,5].

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# Bacterial Pathogens Associated with Community-acquired Pneumonia

We read with much interest the recent article in *Indian Pediatrics* by Das, *et al*. [1], and have the following comments to offer:

1. The authors mention that "in cases of *S. pneumoniae*, *K. pneumoniae* and *S. aureus*, all cases detected by PCR analysis of the respiratory samples were also detected by culture." Authors have not provided the number or proportion of cases detected by PCR and culture. The bacterial load and antibiotic sensitivity of the culture positive cases would have contributed to the existing knowledge.
2. The use of oropharyngeal aspirate as the sample for isolation of bacterial pathogens associated with community acquired pneumonia (CAP) raises many questions. This is again highlighted by the isolation of organism like *Acinetobacter* and *Citrobacter* species from CAP cases. The value of isolating bacterial organisms that are frequently detected in the upper airways of children (eg, *Streptococcus pneumoniae, Haemophilus influenzae, Staphylococcus aureus*) are questionable. Nevertheless, had the authors provided the serotypes of the pneumococcal isolates, the presence of serotypes that are rarely found in the upper respiratory tract but are well recognized causes of invasive disease (eg, serotype 1), may have been highly predictive of pneumococcal pneumonia [2].
3. There is no mention whether the children had any pre-existing respiratory morbidity, as chronic respiratory diseases would significantly influence the bacterial flora.
4. The authors did not mention whether the children received antibiotics prior to sampling. Stralin, *et al*. [3] demonstrated that use of antibiotics decreased the yield of culture for *S. pneumoniae* significantly compared to PCR.
5. The conjugate *H. influenzae* vaccine is known to decrease the nasopharyngeal carriage of the organism [4], and many of these children might have received this vaccine as per latest National Immunization Schedule. As all the *H. influenzae* isolates were 'non type b', the data on H influenzae immunization status of the children would have been interesting.
6. Nasopharyngeal carriage of *S. pneumoniae* has been used as a surrogate marker for invasive disease in children with pneumonia [5]. The data on treatment received by the children and their outcome would have enlightened the readers about the clinical usefulness of the isolates in the absence of a positive blood culture.

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# Bacterial Pathogens Associated with Community-acquired Pneumonia : Author's Reply

We offer the following comments in response:

1. All the cases of *S. pneumoniae (n=32), K. pneumoniae (n=*23*)* and *S. aureus (n=15)* were detected by both PCR analysis and culture of the respiratory samples. In conventional PCR, bacterial load estimation was not possible. Antibiotic sensitivity testing was not intended in this study.
2. The limitation of oro-pharyngeal swab sampling was already mentioned in the article. Although organisms like *Streptococcus pneumoniae, Haemophilus influenzae, Staphylococcus aureus* are frequently detected in the upper airways, these organisms were considered as causative agents only when these were isolated in significant count with the absence of growth of other commensal organisms.

3. No child had any chronic respiratory disease.
4. Sample collection of the cases was done following admission to the hospital and initiation of the investigation procedure. The first dose of empirical antibiotic therapy was already administered.
5. We admit the limitation of missing immunization history against *Haemophilus influenzae* B.
6. Follow-up of the patients as regarding the treatment course was not carried out in this study.

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# Rubinstein-Taybi Syndrome with Psychosis

Rubinstein-Taybi syndrome is characterized by a broad thumb and bulbous hallux, short stature, intellectual disability and distinctive facial features [1]. It is a rare neuro-developmental disorder with a reported prevalence of 1 in 1,25,000 births [2]. Psychosis in RTS is highly infrequent with only a few scattered case reports [3]. A comprehensive literature search yielded only one case report of non-affective psychosis [4].

A 15-year-old girl was admitted to our department with spells of irritability and aggression for last 20 days. These episodes were accompanied by abnormal behavior like singing aloud and pacing. She appeared fearful, and was clinging to her mother. Upon detailed evaluation, there were no well- formed delusions and no clear-cut affective component could be distinguished. Therefore, a diagnosis of non-affective psychosis (Hallucinatory psychosis; ICD F28) was made. Behavioral problems were rated on the Brief Psychiatric Rating scale for Children (BPRS-C) on admission and 6 weeks later on follow-up.

Clinical examination showed short stature, with a height of 129 cm (below 50$^{th}$ percentile). The thumbs were broad and flattened, as were the terminal phalanges of the other digits. The great toes were short and bulbous. There was microcephaly and typical facies, with a low hairline, hypertelorism, bushy eyebrows, broad nose and open mouth. Thoraco-lumbar scoliosis was noted. Muscle tone was low globally. Multiple keloids were present over the left scapular region and popliteal regions of both knees. Findings were consistent with a diagnosis of Rubinstein-Taybi syndrome.

Investigations revealed normocytic hypochromic anemia. MRI spine showed thoraco-lumbar scoliosis and decreased vertebral height. Intelligence Quotient on Binet-Kamat test gave a score of 57, indicating mild intellectual disability. Cytogenetic analysis by Giemsa showed a normal karyotype (46, XX). The patient was started on Quetiapine and recorded a reduction of more than 50% in BPRS-C scores at 6 weeks on a dose of 50 mg, indicating significant response to therapy.

The association of psychosis with Rubinstein-Taybi syndrome is rare and only a handful of cases have been reported in literature. A novel study from Japan determined that variation in the promoter region of the same *CREB* gene may modify gene expression and contribute to schizophrenic psychosis [5]. The rare co-occurrence of psychosis in this syndrome thus opens up a narrow window of opportunity to identify the common genetic changes that result in this combined phenotypic manifestation. This, in turn, may generate fresh insight into the genetic markers of childhood psychosis.

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# Early Presentation of Cherubism

Cherubism is a rare genetic disorder with approximately 300 cases reported worldwide. The disorder typically begins in children at ages of 2-7 years affecting males and females with equal frequency [1]. The lesions usually first appear symmetrically in the angle of mandible; rarely involvement of condyles and zygomatic arches has been reported. Lesions are limited to the jaws, and in most cases begin to regress with the onset of puberty. Respiratory problems due to backward displacement of tongue or obliteration of the nasal airway may manifest as upper airway obstruction. Extracranial involvement is extremely rare. Biochemistry is usually normal in these patients [2].

A 9-month-old girl presented with progressive enlargement of the facial bones first noticed at 3 months of age. The enlargement was gradual, involving the maxilla and the mandible bilaterally initially (***Fig.* 1**); followed by development of palpable firm to hard lesions over affected bones without any pressure symptoms. She was referred with a probable diagnosis of fibrous dysplasia. CT scan revealed symmetrical enlargement of mandibles involving the body, ramus, coronoid and condylar processes with loss of normal trabecular pattern and ground glass opacity in involved bones (***Fig.* 1**). Maxilla, sphenoid wings, body and pterygoid plates showed similar changes. A bone scan revealed overgrowth of mandible with increased uptake and no other significant bony abnormality. A biopsy was advised but the parents refused for the same.

Grading systems for cherubism have been suggested to describe location and severity of lesions. There are no distinguishable histological lesions specific for cherubism. The disease usually occurs due to dominant mutations on *SH3BP2* gene located on chromosome 4p16.3 [3,4]. The differential diagnoses include brown tumor of hyperparathyroidism, giant cell lesions, fibrous dysplasia, aneurysmal bone cyst and the hyperparathyroidism-jaw tumor syndrome.

Follow-up every 2 to 5 years is advisable after the disease becomes quiescent. Surgical intervention is indicated when aesthetic or functional concerns arise.

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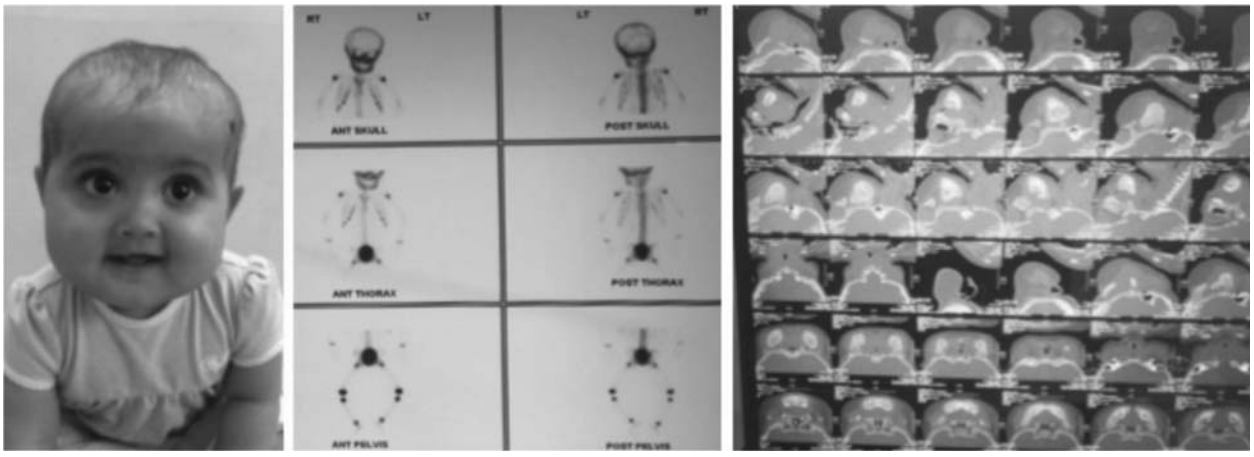


(*a*) (*b*) (*c*)

***FIG.*1** (*a) Typical faces of child showing fullness of maxilla and mandible with upward slanting of eyes and the sclera visible below the irises;* (*b*) *Bone scan demonstrating increased uptake in the mandible and maxilla with no other abnormality;* (*c*) *CT scan of the child showing symmetrical enlargement of the mandibles.*

# Middle East Respiratory Syndrome Coronavirus in Children

Middle East respiratory syndrome coronavirus (MERS-CoV) is one of the recently encountered viral diseases causing pulmonary infection in children. Till date, about 16 pediatric cases are reported in the literature [1,2]. This new coronavirus belongs to lineage C of the genus Beta coronavirus and is genetically closely related to coronaviruses from various bat species in Africa, Middle East and Eurasia [3]. In 2012, the novel human coronavirus was identified in two adult patients with severe respiratory disease in Saudi Arabia.

MERS-Cov usually spreads by droplet inhalation, and the case fatality is very high in adults. In contrast, the disease is usually mild in children [4]. Fever with cough the predominant clinical symptom in the majority of affected children, with occasional rapid deterioration and increasing oxygen requirements, requiring mechanical ventilation and ECMO (Extracorporeal Membrane oxygenation). Acute respiratory illness has been noted in only four cases with a fatal outcome following multi-organ failure [1,2,4]. All these four cases involving pediatric patients were associated with comorbidities such as nephrotic syndrome, Down syndrome, craniopharyngioma and right ventricular tumor. The diagnosis of MERS-Cov is currently established by positive real-time reverse-transcriptase polymerase chain reaction (rRT-PCR) in deep nasopharyngeal secrations.

On imaging studies, the MERS- CoV pneumonia has a radiographic appearance that mimics other more common pulmonary viral infections. Ground-glass opacity (66%) was the most commonly encountered abnormality in adults followed by consolidation (18%) [2]. In children, fine reticular pattern interstitial inflammation may be seen [4]. In acute respiratory illness, the milder lung disease may rapidly progress into diffuse bilateral ground glass opacities mixed with air space consolidation. Pleural effusion or chest *X*-ray was noted in only one of these cases [5]. Although, the extent of lung involvement can be better estimated with computed tomography, sequential chest radiographs have an additional advantage of estimation of the chest radiographic score and the chest radiographic deterioration score with an acceptable dose of radiation exposure to the individuals [2]. Due to its nonspecific clinical symptoms, the accurate and timely diagnosis of MERS-CoV in children can be challenging. However, heightened clinical suspicion in children with underlying risk factors living in endemic areas in conjunction with improved understanding of imaging findings have a great potential for optimal pediatric patient care.

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