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# Kawasaki disease – tissue scurvy misdiagnosed as non-accidental injury

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**Abstract:** Kawasaki Disease is an illness of unknown aetiology,occurs worldwide,and mainly affects Japanese or Asian children under 5 years of age. One feature of the disease, extensive bruising, suggested the possibility of Vitamin C deficiency since it is one of the main causes of bruises in children. Vitamin C deficiency may present in the form of "Tissue Scurvy" – an autoimmune disorder in which there is an abundance of Vitamin C in the body(unlike the Seafarer Scurvy of yesteryear)but it is inhibited from entering the cell to perform its functions because of the lack of insulin which is essential for the transfer of Vitamin C into the cell.Here it is shown a child presenting with the signs and symptoms of Kawasaki Disease was found to have hyperglycaemia, implying insulin deficiency.It is concluded that Kawasaki Disease is an autoimmune disorderfollowing antigenic stimulation in a genetically susceptible child.

**Keywords:** Kawasaki Disease, Non-accidental Injury, Tissue Scurvy

## 1. Introduction

"The incidence of Kawasaki Disease has continued to grow. Why? Why can't we stop this disease? The reason, unfortunately, is that its cause is not known"[1]

Kawasaki Disease, first described by Tomisaku Kawasaki, is an illness affecting children under 5 years, commonly between 6 and 12 months of age, who usually present with fever, redness of the eyes, blotchy red skin lesions, changes in the mouth and cracked lips, swelling of the hands and feet and lymph node enlargement in the neck.

The disorder appears world-wide and Asian, especially Japanese; children are at the highest risk. It is an acute febrile (necrotizing) vasculitis frequently involving the coronary arteries causing coronary thrombosis a leading cause of death in the condition.It has replaced acute rheumatic fever as the leading cause of acquired heart disease in children in the United States and Japan[2].

It was suggested that Vitamin C has a role in the pathogenesis of Kawasaki Disease because Vitamin C maintains the integrity of the vascular endothelium,increasedthe synthesis and deposition of type 1V collagen in the basement membrane, stimulated endothelialproliferation, inhibited apoptosis, scavenged radical species and may play a role in preventing endothelial dysfunction[3].

Cunningham et al[4]; demonstrated reduced mononuclear leucocyte ascorbic acid content in adults with insulin dependent diabetes mellitus consuming adequate dietary Vitamin C, an observation which eventually lead to the concept of Tissue Scurvy.

Tissue Scurvy, as distinct from Seafarer Scurvy, a nutritional deficiency, is a condition in which there is an abundance of Vitamin C in the body but it is inhibited from entering the cellto perform its functions because of the lack of insulin which is essential for the transfer of Vitamin C into the cell.

Normally insulin binds to its receptor on the cell surface and initiates a chain of events that leads to the insertion in the plasma membrane of a transmembrane glucose transporter called GLUT 4 which facilitates the transport of glucose and other nutrients into the cell. It is the failure of this process which prevents the entry of Vitamin C into the cell and causes "tissue scurvy" which affects every cell in the body to a greater or lesser degree[5].

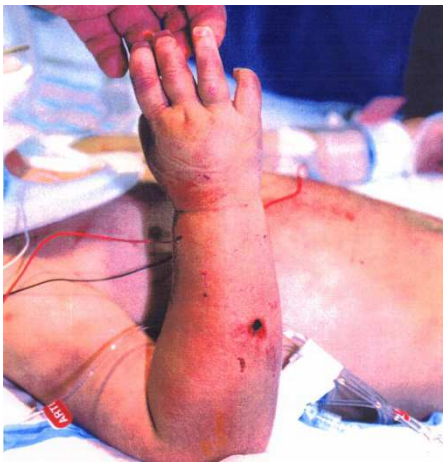
Tissue Scurvy, when it involves the cells of the liver leads to under carboxylation of the coagulation factors II, VII, IX and X and of osteocalcin which could result in bruises, hemorrhages and fractures[6].

## 2. Method

To determine whether a similar situation was the cause of Kawasaki Disease the clinical and laboratory investigations of a child alleged to have been physically abused were re-examined. It was found the child had the clinical features of Kawasaki Disease and his Blood Tests showed evidence of hyperglycaemia proving a deficiency of Insulin and thereby establishing Tissue Scurvy as the cause of his physical signs and symptoms.

### *2.1. Case Report*

A three year old boy was brought to the hospital Emergency Department by his mother with the complaint that he was not "breathing right." The child had been left in the care of the mother's partner.



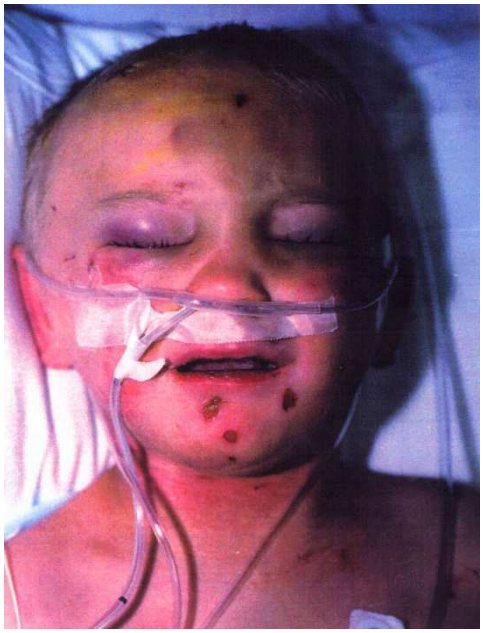
***Picture 1.*** *Swelling of hand and arms*

The child was being punished and given "time out" by being made to stand in the corner for some minor dispute with his siblings. While there he suddenly collapsed, fell to the ground and stopped breathing.

The carer immediately commenced CPR with marginal success in that the child started breathing spontaneously but irregularly and became limp and unresponsive.

The mother was called and together they went to the hospital where the attending doctor noted the child was grey, limp, unconscious and unresponsive, hemiparetic with "ecchymoses from head to toe" and "declared a Trauma, Level 1." It was also noted that "his hands were swollen into balls as if they had been crushed" and the feet had round reddened areas.

Further examination, according to the admitting doctor, showed "multiple bruises on the abdomen, back, face, legs? bite marks, ?ligature marks around neck" and the bruises were of "varying ages". Some of the bruises had a central pallor and some "resembled cigarette burns".

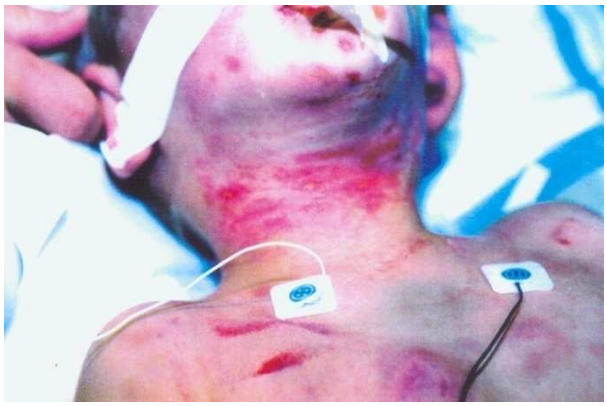


***Picture 2.*** *Multiple bruises on the face*

A Consultant Paediatrician noted:-

1.Lichenified rash on neck
2.Laceration left side forehead
3.Bruises on both sides of the face
4.Laceration right cheek.
5. Purple spots on the chest.
6.Linear bruises on the left shoulder
7.Eschars both arms and back and right side of the abdomen.
8.Finger tip marks on the right arm
9. Scattered bruises on both legs
10. Red marks on the scrotum
11. Red mark on the left hip
12.Swelling of both hands and arms.

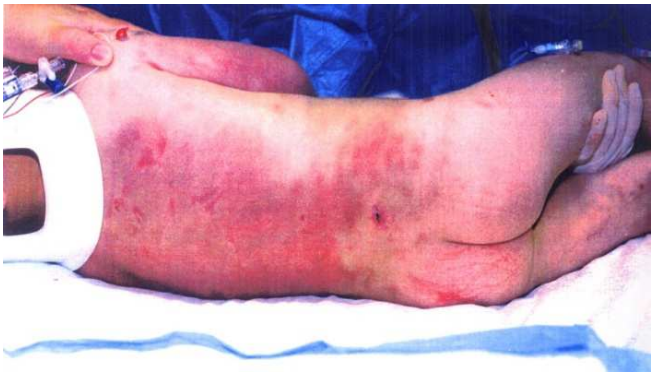
No explanation could be offered for the bruises by the mother or boyfriend other than the child bruised easily and was continually falling off his bicycle, having refused to use "trainer wheels". Furthermore his older siblings play rough and "kick him in the groin" sometimes. His mother had noticed the swelling of the hands some days earlier and the boyfriend, thinking the child had developed a rash on his neck, applied some cream. Dried "cracked lips" also were noted and cream applied. The child had been vomiting recently and complained of a headache for which he was given Tylenol.



***Picture 3.*** *Polymorphous skin lesions*

A MRI Brain Scan "showed small bilateral occipital and tentorial subdural haemorrhages somewhat larger on the right than the left side. A small left frontal subdural effusion. A moderate size left temporal haemorrhagic contusion and multiple areas of abnormal diffusion and FLAIR signal involving the left frontal, occipital and temporal lobes as well as the right parietal and occipital cortex, the superior medial right frontal areas of cortex level cerebellar hemispheres. These diffusion and FLAIR signal abnormalities likely represent underlying cortical oedema".

"There was blood around the bowel and in the retrosternal space".



***Picture 4.*** *Scattered bruises over the back and legs*

Significant Laboratory Investigations on admission were:-

***2.1.1. Evidence of Autoimmune Beta Cell Dysfunction***

1. Glucose 141 mg/dLNR 60 – 115 mg/dL
2. ALT 177 U/L NR 7 – 90 U/L
3. AST 187 U/L NR 5 – 60 U/L
4. GGPT 91 U/L NR 8 – 78 U/L
5. Albumin 2.8 g/L NR 3.4 – 5.1 g/L

***2.1.2. Evidence of Acidosis***

Blood pH 7.29 NR 7.32 – 7.42

***2.1.3. Evidence of Anaemia***

1. Hct 32.6 % NR 34 – 40 %
2. Hgb 10.9 g/dl NR 11.5 – 13.5 g/dl

***2.1.4. Evidence of a Coagulation Disorder***

1.Prothrombin Time 11.7 secs NR 10.1 –11.5
2.PartialThromboplastin Time 23 secs NR 26 - 38
3.Platelet count 240 K/cc mm NR 140 – 440
4.Fibrinogen 427 mg/dl NR 200 – 466

## 3. Discussion

The Evidence of Kawasaki Disease is:-

Classically Kawasaki Disease is suggested by FEVER of more than 5 days duration and at least four of the five following conditions:-

1. Non-purulent conjunctivitis. Non-exudative bulbar conjunctival injection
2. Oropharyngeal changes (injected pharynx, strawberry tongue, fissured lips)
3. Polymorphous skin lesions
4. Cervical Lymph node enlargement
5. Extremity changes (erythema of palms and soles, edema of the hands and feet, periungual desquamation)

In addition elevation of liver enzyme values, usually two to threefold is common in Kawasaki Disease according to Leung et al.[7]

Atypical Kawasaki Disease presents with some of these features, not necessarily appearing at the same time.

In this child the polymorphous skin lesions (described, erroneously, as bites, cigarette burns, finger marks, eschars and strangulation marks), marked oedema of the hands and arms, erythema of the feet, fissured lips, vomiting and a two-fold elevation of liver enzymes leaves little doubt regarding the diagnosis.

A low blood pH, such as one finds here, is indicative of a change in the metabolism of the body resulting from the non-availability of oxygen as seen in either cardiac or respiratory arrest.

This result supports and is entirely consistent with the history given by the carer that the child suddenly stopped breathing went grey and limp and suffered what is an Apparent Life Threatening Event (ALTE).

The alacrity with which the admitting doctor declared "Trauma Level 1" coloured subsequent clinical and investigative procedures. The possibility of Kawasaki Disease was never raised and the Anti-neutrophil cytoplasmic antibody (ANCA) test which is sometimes positive in this type of vasculitis was never done.

However the presence of hyperglycaemia is clear evidence of Insulin deficiency provoking the onset of Tissue Scurvy. The coronary vessels and myocardium should have been thoroughly investigated as they are a major cause of death in Kawasaki Disease.

## 4. Conclusion

Leung and Meissner[7] suggested there are "Many Faces of Kawasaki Syndrome" and the many dysfunctional tissues created by Tissue Scurvy would certainly explain those many faces.A prominent role of vitamin C in

maintaining the integrity of the vascular system is clearly dysfunctional in this child.

Vitamin C is required for the synthesis of collagen, the intercellular "cement" substance which gives structure to muscles, vascular tissues, bones, tendons and ligaments. In addition, vitamin C contributes to preventing haemorrhaging and bleeding. It also improves the absorption of iron from the diet and helps prevent anemia seen in this child.Another important function of Vitamin C is its role in immunity where it functions as an anti-allergic agent since it is a natural antihistamine. It both prevents histamine release and increases the detoxification of histamine.It is the failure of this anti-allergic function which initiates all the features of Kawasaki Disease establishing it as an Autoimmune Disorder

## Acknowledgements

I wish to thank the family for permission to use the photographs demonstrated here and I wish to thank my wife for helpful insights.

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