

原文題目(出處)：	Radicular cyst in a patient with untreated Wiskott–Aldrich syndrome: A case report. Oral Sci Int 2013;10:40-3
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內文：

### I. Abstract

Wiskott–Aldrich syndrome (WAS) is a condition with variable expression, which causes persistent thrombocytopenia and, in its complete form, also causes small platelets and humoral immunodeficiency. A 14-year-old boy, diagnosed with WAS but never treated, presented with symptoms of heart and renal failure. His right buccal region was swollen and his right first molar showed a cyst-like image on dental X-ray films. The boy's symptoms were attributed to an infected cyst, greatly aggravated by WAS-related immunodeficiency. The boy was sedated and the affected tooth and cyst were enucleated. Invasive treatment was safely achieved by paying close attention to whole-body management.

### II. Introduction - Wiskott–Aldrich syndrome

1. A congenital X-linked immunodeficiency
2. Characterized by frequent infections, thrombocytopenia with small platelets, eczema, and an increased risk of autoimmune disorders and malignancies
3. In 1994, WAS was linked to mutations in a gene on the short arm of the X chromosome, encoding the WAS protein.
4. Mean survival time of 6.5 years from birth
5. Report a 14-year-old boy with untreated WAS who underwent a successful mandibular cystectomy

### III. Case report

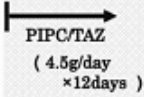

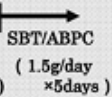
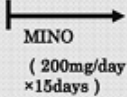
1. General data : A 14-year-old boy
2. Chief complaint : Swelling over R't buccal region  
Caries and PAP of 46
3. Past medical history & family history :  
Wiskott–Aldrich syndrome  
Renal insufficiency  
Bilateral pulmonary edema  
Anemia  
Intellectual disability
4. Alcohol、Cigarettes : unremarkable



**Table 1**  
Changes in perioperative laboratory values.

Days of hospitalization	1 (ER)	13 (Pediatric ward)	35 (Preoperative day)	36 (Post platelet transfusion)	37 (POD1)	43 (POD7)	56 (POD24)
WBC	9.3	4.5	4	2.8	3.7	4.1	4.9
RBC	99	178	186	170	182	212	181
Hb	2.7	5.2	5.3	4.9	5.1	5.9	4.8
Pit	4.7	1	1.3	6.8	6.9	1.4	1.4
BUN	68	33	28		26	38	58
Cr	7.5	4.8	2.5		2.8	3.8	5.2
K	6.4	5.2	4		4.8	5.2	4.5
CRP	12	0.2	0.6		0.7	0.2	1.7

5. Intra-oral examination :  
Caries of tooth 46  
Impaction tooth of 18, 28, 38, 48  
A 2x2.5cm RL region of 46 apical region
6. Extra-oral examination :  
Swelling over R't buccal region
7. Clinical differential diagnosis : unavailable
8. Histological examination : unavailable
9. Histologic Diagnosis: unavailable
10. Treatment :  
Antibiotics for infection  
Restricted diet, and limited water intake  
Cystectomy

Days of hospitalization	<i>ER</i>			<i>pediatric ward</i>			Operation		Discharge			
	1	4	12	17	20	29	33	35	49	54	56	
Postoperative day							1	7			24	
a course of antibiotics	 PIPC/TAZ ( 4.5g/day ×12days )						 CTX ( 1.0g/day ×5days )		 SBT/ABPC ( 1.5g/day ×5days )		 MINO ( 200mg/day ×15days )	
erythrocyte transfusion (ml)	400	400				400						
platelet transfusion (ml)				200	200		200	200			200 200	



**IV. Discussion**

1. Limit water intake and maintain strict dietary restrictions in order to correct electrolyte imbalances and prevent aggravation of renal dysfunction.
2. Patient has a blood platelet count of 10,000–20,000/ $\mu$ L, an operation such as a tooth extraction can normally be performed safely with local hemostasis.
3. Vasoconstrictors, mixed with local anesthetics making it safe to use the

anesthetic agent for an extended duration and reducing the risk of hemorrhage, but may induce necrosis when used in anemic tissue

4. Mepivacaine does not have a vasodilating effect, helped to minimize the risk of delayed wound healing due to local anemia.
5. Propofol is the sedative of choice for these procedures because it is metabolized mainly by the liver

題號	題目
1	下面關於根尖囊腫(radicular cyst), 何者錯誤? (A) 常因根尖表皮被發炎刺激所產生 (B) 表面由分層鱗狀上皮(stratified squamous epithilium)覆蓋 (C) 囊腫內充滿液體及細胞殘骸, (D) 不會發生在乳齒中
答案(D)	出處: oral and maxillofacial pathology 3rd ed
題號	題目
2	下面關於根尖囊腫(radicular cyst), 何者錯誤? (A) 囊腫發源牙齒會失去活性 (B) 患者常有脹痛感 (C) 需進行根管治療 (D) 常因拔牙後傷口未清除乾淨導致
答案(B)	出處: oral and maxillofacial pathology 3rd ed